OPEN MYELOMENINGOCELE

A five year review of 200 consecutive closures

by

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This work is dedicated to my wife and daughters for their unfailing support of me and to many parents for their unfailing support of their handicapped children.

James Lister

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The cases studied in this thesis were referred to the Paediatric Surgical Unit at the Children's Hospital in Sheffield and all operations on the back, on hydrocephalus, and on the renal tract were performed by members of the surgical team. Patients were in the care of my paediatric surgical colleague, Mr. R.B. Zachary, and myself. It is clear that the amount of work involved in treating these children must also involve many others; Mr. W.J.W. Sharrard is the orthopoedic surgeon who gave unstinting care throughout, Dr. John Lorber was the paediatrician from whom we continually sought advice. Dr. R.K. Levick, Radiologist, and Dr. J.L. Emery, Pathologist, contributed greatly to the management of these children.

The research work which is quoted from time to time, particularly in regard to the urinary tract, was undertaken by a series of Research Fellows, Mr. R.C.M. Cook, Dr. E. Pekarovic and Mr. R.E. Cudmore. The research work was carried out in the Wolfson Building for Research into Congenital Deformities, a part of the Department of Child Health of the University of Sheffield, supported partly by charitable donations and administered by a Research Committee consisting of Dr. J.L. Emery, Dr. R.K. Levick, Dr. J. Lorber, Mr. W.J.W. Sharrard, Mr. R.B. Zachary and myself.

I have quoted freely from my colleagues but the review of the material has been entirely my own work as has, of course, the discussion. The conclusions drawn are personal ones which are, nevertheless, shared by my surgical colleagues. Dr. Lorber's contrasting views have been quoted in the thesis.

I would particularly like to acknowledge the work of Mr. R.B. Zachary who pioneered the aggressive approach to myelomeningocele and whose many previous years of experience, which would have disheartened most men, eventually brought their dividends when the Holter valve became available for ventriculo-cardiac shunts.

INDEX

INTRODUCTION

Paediatric surgery has been defined as the surgical care of the newborn, the infant and the child. The specialty has grown in the United Kingdom as in other parts of Europe and the world in a fairly close relationship to improving social standards and medical care. Although in Edinburgh and Glasgow paediatric surgeons were established as specialists from the early 1920s, it was not until 1950 in England and Wales that the number of paediatric surgeons exceeded 4, and in 1971 there are a total of 34 surgeons in the United Kingdom confining their practice to paediatric surgery. Whilst few paediatric surgeons would claim that the care of a child with a condition such as appendicitis demands skill and expertise beyond that available to a general surgeon, paediatric surgeons, and indeed most general surgeons, would agree that surgery in the newborn child and in the infant suffering from the less common types of congenital abnormalities demands not only special surgical experience but more importantly a team of medical, nursing and technical specialists with sophisticated experience and equipment only available in a special unit.

At the turn of the century of every 1000 liveborn babies 140 died within the first year of life; the main causes of this infant mortality rate were prematurity, birth injury, respiratory difficulties and infections, and congenital abnormalities. The infant mortality rate in 1969 was 18 and this enormous reduction has been achieved by improvements in social conditions affecting the pregnant mother, together with improvements in obstetric care and the tremendously successful care of the newborn child, developed during the past thirty years. As control of the other lethal factors has been achieved the infant mortality from congenital abnormalities, remaining unchanged at 4 per 1000, has assumed the role of major killer, and efforts have been directed at reducing it. For example, in the Children's Hospital in Sheffield in the late 1940s some 30 newborn children were operated on each year, the number operated on now is more than 10 times as many.

Not all congenital abnormalities are incompatible with life and whilst the correction of atresias of the alimentary tract or diaphragmatic hernias are dramatic life saving procedures, congenital abnormalities of the cardio-vascular and central nervous systems are far more common and at the same time less

immediately lethal. The correction of congenital heart disease during the first three months of life has already altered the outlook to this type of abnormality and will continue to do so for many years. Congenital abnormalities of the central nervous system, however, are far less dramatic in the toll they take of the child in his first year of life and yet they are the commonest group seen at birth. The most common lesion is a myelomeningocele with exposed spinal cord and consequent neuropathic changes in the lower limbs, pelvic floor and bladder and bowel sphincters. Until 1957 no satisfactory treatment was available for the control of the hydrocephalus which almost universally was an associated abnormality with the severe lesions; there was little encouragement, therefore, for an attack on the back lesion when it was almost certain that the child would die of progressive hydrocephalus within a year or two. However, the introduction of a valve system of ventriculo-cardiac drainage produced a method for control of progressive hydrocephalus and it soon became clear that an early covering of the exposed spinal cord on the back would preserve such function as already was present in the lower limbs and pelvis, whilst a negative approach leaving the closure to a slow process of granulation and epithelialisation was almost certain to increase the degree of paralysis because of progressive damage to the exposed nerves. In many centres, therefore, an early vigorous attack on the exposed lesion of the back was introduced and as the interest of the paediatric surgeon in these cases became known to obstetricians and paediatricians the number of cases referred for early treatment increased. In an area such as Sheffield where the paediatric surgical unit drains a population of approximately 3 million it could be expected that 120 cases would be referred each year and this number represents almost the entire incidence of the condition in the region. The estimated incidence of the condition is between 2 and 3 cases per 1000 live births and with a birthrate of 16 per thousand this would produce 40 cases for each million population in each year.

It is too early as yet to decide whether the early treatment of the backs in these children will effect the long term survival rates in the condition and certainly it seems likely that some children die as a result of the surgical intervention who might not have died had their backs been left to granulate; at the same time there is no doubt that many children live much longer as a result of the early surgical intervention and nor is there any doubt that having undertaken the primary treat-

ment of the back in a child with myelomeningocele the surgeon and his unit must hold themselves responsible for the continuing treatment of the child, whether such treatment is necessary because of complications of the original surgical interference, or whether it is necessary to control the progress of those parts of the disease which were not corrected at the original surgical interference. Thus, having closed the back it is necessary that progressive hydrocephalus should be treated as required, that appropriate orthopaedic procedures should be carried out in order to achieve and preserve the child's mobility, and that the upper urinary tract should be protected and preserved from the devastation brought by the combination of urinary obstruction and infection resulting from the neuropathic bladder.

Not all this complicated care can be carried out by the same person who originally closed the back and the general care of these children must depend on a team which will include at least a paediatric surgeon, an orthopaedic surgeon and a paediatrician. The particular interests of individuals will of course influence the number of specialists taking an interest in these children. An indication of the amount of work that the continuing control of these children must bring to a hospital is the fact that the Children's Hospital in Sheffield with an annual intake of 100 to 120 new cases for the past eight years, up to 40 of the total of 200 beds available in the hospital are now occupied by children having treatment for spina bifida. This does not include those children who are being cared for in hospitals nearer their own homes or those in special long term orthopaedic hospitals.

The condition thus places a heavy load on the hospital and its staff. Clearly as medical and surgical care develops and efficiency of treatment improves, the case material in a hospital will change; for instance at one time tuberculosis of bones and joints accounted for a very large number of those children undergoing surgery in a children's hospital and today it is practically unknown in the United Kingdom. Similarly, infections such as osteomyelitis and empyema which at one time formed a large proportion of the work in the children's hospital are now very rare. These conditions have been excluded because they have been cured by advances in medical and surgical treatment. The surgical treatment of congenital abnormalities, however, and particularly the surgical treatment of myelomeningocele often does not cure but only controls the condition, and in fact in these conditions the paediatric surgeon frequently produces a new population which will demand

repeated and often prolonged hospitalization. It would seem necessary, therefore, that those who are concerned in the treatment of congenital abnormalities should consider the effects of that treatment. For the physician some consideration must be given to the quality of survival achieved as a result of extensive and continuing courses of treatment. For the administrator and those who contribute to the financing of the health services consideration should presumably be given to the value of the results achieved; the financial resources of the health services are not unlimited so there must be some competition for those resources and consequently one group of patients may suffer because of the concentration of funds on another group. From a purely business outlook of cost and return there would be little difficulty in deciding that the funds of the health services should be concentrated on those patients who will give the most productive return to the country for the money that the country has spent on them.

Fortunately for the medical profession our decisions are not governed entirely by financial considerations and the purpose of this thesis is to consider the results achieved in the treatment of myelomeningocele and to discuss those ways in which these results might be improved.

HISTORICAL

Spina Bifida

It is perhaps strange that congenital abnormalities so obvious as spina bifida and myelomeningocele should have been so infrequently reported in early writings. Cooper in his Dictionary of Practical Surgery and Encyclopaedia of Surgical Science states that the Arabians were the first to treat the disease but gives no reference and Denuce quotes a reference to the condition in Hippocrates. It was Nicholas Tulp, however, who first coined the phrase "Spina Bifida" in 1652 and from that time on the condition was more frequently mentioned by medical writers. There was little evidence of interest in the treatment of the disease until well into the 19th century; it was accepted that this was a condition almost never seen in adults, a condition that was likely to prove fatal before the age of three years and more often to produce sudden death after the child had lingered for only a few months from birth. Death usually followed operations upon the tumour and Tulp himself reported instant death as a result of an attempt at surgical correction.

In 1834 P.S.K. Newbigging, a son of a President of the Royal College of Surgeons of Edinburgh and later himself President of that College, chose spina bifida as the subject of his probationary essay for admission to Fellowship of the College. Newbigging discussed the aetiology of the condition believing it to be an arrest in development as there were so many associated anomalies occurring with it; he discussed very fully the pathology of the condition, and he recommended a bold approach in its treatment. He reported three successfully treated cases, two by puncture of the cystic lesion and one by excision of the lesion. In view of the uniformly fatal results he felt that this approach was justifiable and much preferable to palliative measures using astringent lotions, mild plasters, or bandages with lead plates (as recommended by Astley Cooper). Newbigging, however, admitted that "some modern authors disapprove in strong terms of using any treatment whatsoever ..... prolonging for days or weeks the vegative existence in a being that is not viable".

In 1877 James Morton published a monograph reviewing some 50 cases of spina bifida and myelomeningocele treated by a method of injection. Ten years

later a second edition was published bringing the total number of cases treated to 71. Morton was a surgeon in the Glasgow Royal Infirmary who was born in 1820 and died while still working in 1899. He was Professor of Materia Medica in the Andersonian Medical School for 33 years from 1855 until 1888, yet it was as a surgeon that he made his mark, and in the development of a new method of treatment of spina bifida. His monograph includes a very detailed description of the pathology of the condition and it was perhaps because of this and because of the review of previous experiences in treatment that Morton's work was taken so seriously. He reported 46 cases treated by puncture or incision of whom 30 died, 12 recovered and 4 were unrelieved or unknown; he noted that 14 of the children had died of meningitis and this is the first mention of meningitis as a complication of the condition. He believed that ligature and excision of the sac was only applicable to a limited number of small pedicled lesions since only in these cases was there no risk of damaging nerves. Howson in 1884 had reported a lumbar myelomeningocele treated by the insertion of Southey's tubes and the injection of tincture of iodine: this had led to an inflammatory reaction in the sac and allowed later excision of the tumour. Morton developed the method using an iodo-glycerine solution containing 10 grains of iodine and 30 grains of iodide of potassium dissolved in 1 ounce of glycerine. A variable amount of this solution was injected into the meningocele after aspirating some fluid and light pressure was maintained to avoid leakage. Morton believed that this treatment was indicated in all cases where death was to be feared from rupture of the sac and he believed that the time of operation should be within a month of birth provided other conditions were favourable.

Morton's experience was extensive and he claimed 54 cures out of 67 cases, though he admitted that in his own hands the mortality rate was something over 20%. Cure probably referred to disappearance of the swelling rather than prolonged life but these results were sufficiently impressive for the Clinical Society of London to set up a sub-committee to investigate the treatment of spina bifida by the injection of iodo-glycerine solution, and this committee having considered various other types of treatment concluded in 1885 that the injection treatment was justifiably favoured and widely used. They recommended that the best results were to be hoped for in children who had reached the age of two months, in

whom there was no paralysis or hydrocephalus and in whom the sac was covered by healthy skin.

In 1900 James H. Nichol, another Glasgow surgeon, condemned the injection method of treatment and recommended open operation. Nichol was one of the first surgeons in the Children's Hospital in Glasgow and later surgeon to the Western Infirmary. He was one of the earliest to describe and operate on congenital hypertrophic pyloric stenosis and also for cranial depression in infants. Perhaps because of his interest in bacteriology he was much in favour of out-patient operations and developed techniques for the day case treatment of hernia and later on spina bifida. For this type of surgery he depended on nursing, not by the mothers but by an organized body of outdoor visiting nurses. Nichol incised the myelomeningocele sac and scarified it with longitudinal incisions in order to encourage adhesion. He felt the method was a safe one, the operative mortality being no higher than the operative mortality in the radical cure of inguinal hernia and he recommended the operation very early in life. He believed that some of his good results were achieved by avoiding infection of the wound, the child being nursed prone on a pillow on the nurse's knee for a week or longer, during which time the back was constantly exposed to view, so that contamination of the dressing by urine or faeces was impossible. He avoided operating on any case in which the sac was ulcerated or sloughing.

Both Morton's and Nichol's methods encouraged the shrinking of the sac which the Clinical Society of London's Committee had stated was the natural cure but gradually both were dropped in favour of excision of the sac. On the whole in the first four decades of the 20th century it seems that surgery was confined to meningoceles with perhaps a small element of neural tissue in them and, indeed, Brenner in 1938 once-more questioned operating on any child who already had paralysed legs and sphincters, describing it as "an act of very doubtful charity". Stockmeyer in 1925 reviewed 213 cases of spina bifida spread over 62 years from 1862 to 1924. Of these 129 had been operated on and 60 had died, deaths being due to meningitis, hydrocephalus, and "over-reduction of intracranial pressure".

The pathology of the varieties of spina bifida was perhaps confused by these earlier authors. Most of the cases operated on were myelomeningoceles

certainly, that is to say a meningocele sac with some nerve fibres running into it, and occasionally those nerve fibres were exposed on the surface of the sac and indeed might represent part of the spinal cord. Myeloceles with a considerable length of the spinal cord lying exposed on the surface of the back but no meningocele sac were not considered for treatment even though in a few days a sac might have developed. If the lesions were not closed within the first few hours of life and the spinal cord was exposed, that is to say if there was a considerable myelomeningocele, then unless the surgeon saw it very early on the child would be completely paralysed by the time he saw it and he would also often discard that one for treatment. Thus most of the cases treated during these periods were what we would today regard as relatively less severe lesions. The progress now towards the almost immediate closure of the back after the baby is born has depended more on the historical progress in the treatment of the hydrocephalus, which so frequently accompanies spina bifida, than on improvements in the back surgery itself.

Hydrocephalus

Hydrocephalus has been known since time immemorial. It was described by Hippocrates and its association with spina bifida was described by Morgagni. The first Scottish author to write on hydrocephalus was St. Clair in Medical Essays and Observations in 1734, and in the same journal the following yearPaisley reported another case. Robert Whytt who was Professor of Medicine in the University of Edinburgh and also at one time President of the Royal College of Physicians of Edinburgh, is credited with drawing a clear delineation between internal and external hydrocephalus in 1734, but his "observations of dropsy of the ventricles of the brain" did not distinguish between hydrocephalus and what we now know to be meningitis. John Cheyne corrected some of the misinterpretations established by Whytt in his essays on hydrocephalus acutus in 1808 and 1819. Cheyne quoted Quinn of Dublin as demonstrating that inflammatory changes in the meninges preceded the formation of increased amounts of cerebro-spinal fluid. John Cheyne was an interesting man born and educated in Scotland being a third generation General Practitioner in Leith who after some time in the Army eventually resolved on specialism in diseases of children and went to Dublin where he became Professor of the Practice of Physic

to the Royal College of Surgeons of Ireland and later Physician General to the Army in Ireland. He wrote three essays on hydrocephalus acutus including one with L.A. Colis who was Physician and Director to the Institute for the Sick Children of the Poor in Vienna. In this particular paper the authors considered the predisposing causes of hydrocephalus and included the great terror and anxiety in the mother which had been seen in the last weeks of pregnancy during the bombardment of Vienna in 1809, and which had led to the birth of children dying from convulsions and showing intracranial inflammation on post mortem examination.

The treatment of hydrocephalus was certainly attempted from time to time by direct drainage (Fig. 1 is a photograph of a mediaeval woodcut showing direct drainage) and by the injection of irritant substances but even after the work of Dandy and Blackfan (1914) had demonstrated the circulation of the cerebro-spinal fluid, various procedures aimed at draining fluid from the lateral ventricles into various body cavities or into other areas of the cerebro-spinal fluid space or even into the venous circulation were disappointing, particularly in children because of the small size of the spaces available, apart altogether from any difficulties with clotting of the drainage systems. Perhaps the most significant of the attempts at returning cerebro-spinal fluid to the venous circulation was that by Payr in 1908 who advocated passing a tube from the ventricle to the longitudinal sinus or the jugular or common facial vein and used as a tube hardened calves' arteries. This system of drainage was the forerunner of the present-day system introduced by Nulsen and Spitz (1952). Spitz, a neurosurgeon from Philadelphia, used a valve designed by Holter to prevent back flow of blood into the plastic tubing running from the lateral ventricles to the internal jugular vein. Spitz never published his results but the Holter valve has gained world-wide acceptance and the drainage from the lateral ventricles into the Holter valve placed subcutaneously just behind the ear, the tube passing from the lower end of the valve into the internal jugular vein in the neck, is the basis of most ventriculo-cardiac shunts. The second of these was described by Pudenz who used a different type of valve which was situated at the lower end of the catheter going down towards the atrium. (Pudenz 1957).

These two systems of drainage were introduced in 1957 and their success

Figure 1 : Mediaeval woodcut

led to an entirely different attitude being developed towards the treatment of spina bifida and hydrocephalus. Once it became possible to control hydrocephalus it could not be long before a more aggressive attitude towards the treatment of myelomeningocele would also develop. If the child was no longer likely to suffer from progressive brain damage and eventual death due to progressive hydrocephalus, then there was good reason to attack the back lesion and protect him from the progression of his paralysis, which was bound to result from infection and drying out of the exposed nerve tissue on the back.

It is thus since 1957 or 1958 that a new approach has been made to the treatment of myelomeningocele. It was in Sheffield that the value of urgent closure of the back was first realised; it was shown (Sherrard, et al 1963) that if the back lesion were closed and the exposed nervous tissue covered within the first 24 hours of life the almost inevitable progression to total paralysis of the lower limbs was stopped. There was perhaps some impression that not only was progression of the disease prevented but some movement of the lower limbs was regained. This was probably a fallacy. There was, however, no doubt that the end results with regard to movement of the lower limbs were better in those children in whom the back lesion was operated on early.

Having made the decision to attack the primary back lesion the surgeon became committed to continue the total care of the child with his subsequent handicaps. These then are the children of whom about 2000 are born each year in the United Kingdom, who present many problems to the paediatric surgeon and his colleagues and who will make continuing demands on the health and social services of the community.

PATHOLOGY

Spina bifida is a general term applied to incomplete closure of the neural tube. The extent of this failure may vary from spina bifida occulta with complete meninges and complete skin covering but a failure of fusion of only one or two spinous processes to complete myelocele where the whole neural groove remains open and where the child is usually anencephalic and stillborn. Myelomeningocele and meningocele are two intermediate degrees of the abnormality; meningocele is a relatively rare condition which is in fact a skin covered meningeal hernia with a fluid containing protrusion of the meninges but no neural element entering the sac. Myelomeningocele may vary from a relatively rare lesion similar to a pure meningocele but with some nerve fibres coursing through the sac and becoming adherent to its inner surface, to a much more severe and more common lesion which was well described by von Recklinghausen in 1886; there is a median cystic swelling on the back with a rather red central apical area described as the zona vasculo-granulosa representing a flattened cord surrounded by a transparent area called the zona pellucida and representing meninges, and extending peripherally to another circular circumferential area of skin often of abnormal colour called the zona epithelic-serosa. These lesions occur most commonly in the lumbo-sacral region though they may also affect the thoracic region.

It should be emphasised that von Recklinghausen and other earlier writers seemed to see these lesions rather later than we do nowadays: it is uncommon for the lesion to be cystic in the first 24 hours of life though it does become so if not repaired. The zona pellucida is a thin translucent membrane of varying width; in some cases the zona epithelio-serosa reaches almost to the neural plaque (zona vasculosa-granulosa) the zona pellucida is so narrow, and here there may be danger that in separating the neural plaque, either nerve tissue is damaged or a small amount of skin is buried and later forms an inclusion dermoid cyst.

In recent years the histology of the exposed neural plaque and the cord above and below it have been studied more closely (Stark et al 1967, Lendon 1968, Emery et al 1969). A great variation in the number of anterior

horn cells in the region of the lesion has been shown and sometimes the numbers are within the normal range. The cord may be simply opened out yet apparently capable of normal function or there may be evidence of agenesis or intra uterine destruction leading to the congenital absence of nerve supply. Abnormalities of the central canal, splitting of the cord, collections of fatty tissue and the survival of islands of reflexly acting neural tissue all contribute to the bizarre effects of paralysis that occur.

Incidence

In the United Kingdom the incidence of spina bifida excluding spina bifida occulta is estimated at 2 per 1000 live births. (Lister 1969). There are considerable geographical variations which may be relative to the genetic component of the aetiological factors. (Laurence 1969).

Aetiology

The study of spontaneously aborted human embryos has shown that closure defects of the neural tube can occur in very young stages just after the time of normal closure of the tube (Kallen 1968) and it seems likely that myelomeningocele is a failure of development rather than a breakdown of a neural tube that had already closed. The latter theory that the already formed neural canal was broken down by pressure from within was that put forward by Morgagni and there are still some people who believe that a hydromyelic neural tube might indeed produce this abnormality. The complicated interactions between the developing central nervous system and the underlying and surrounding mesodermal structures can be quite easily upset in animal experiments, particularly by the use of trypan blue in pregnant rats (Lendon 1968). Only Aminopterin has been shown to produce malformations in the neural tube in man (Thiersch 1952). It is believed that there is an inherited predisposition to the development of myelomeningocele and indeed all degrees of dysraphism, with environmental trigger mechanisms of social class, seasonal and secular variations, parity, maternal age and probably many others (Laurence 1969).

Associated Abnormalities

In almost every case there is a degree of hydrocephalus and this is usually associated with the Arnold-Chiari malformation in which the cerebellum

and the medulla are displaced downwards through the foramen magnum. It has been long accepted that this deformity was brought about by tethering of the spinal cord at the level of the back lesion and consequent displacement of the hind brain in response to the growth differential between vertebral column and spinal cord, normally allowed for by the spinal cord sliding up inside the neural canal. Some doubt has been cast on whether any growth differential exists at all; Barry et al (1957) suggested hypoplasia of the posterior fossa forced its normal contents through the foramen magnum. Whatever the aetiology, the Arnold-Chiari malformation is present in many cases and is believed to plug the foramen magnum so that cerebrospinal fluid cannot pass back into the cranium though it can still pass out of the fourth ventricle into the spinal canal. However, hydrocephalus, apart from the post meningitic variety, can occur in spina bifida without the Arnold-Chiari malformation; and posterior fossa decompression procedures have not been very successful in relieving the hydrocephalus. These two facts lend credence to the alternative theory that the hydrocephalus is a primary associated abnormality and is the cause, not the effect, of the Arnold-Chiari malformation.

That there is an increased incidence of associated abnormalities in spina bifida apart altogether from the hydrocephalus does suggest a general insult to the foetus. Of particular importance is the increased incidence of primary renal abnormalities unassociated with the acquired renal disease resulting from neuropathic urinary obstruction (Roberts 1961, Emergy 1969). But other alimentary anomalies occur and were seen in the series studied.

Clinical Picture

The assessment of the newlyborn child with myelomeningocele may be difficult. The extensive thoraco-lumbo-sacral lesion will be accompanied by almost complete paralysis of the lower limbs, a patulous anus indicating paralysis of the pelvic floor, possibly dribbling urinary incontinence and almost certainly evidence of hydrocephalus in an abnormally large head circumference and an open lambdoid suture. In the meningocele with only limited neural involvement there may be very little evidence of paralysis and

indeed the suspicion of whether the lesion is a meningocele or a myelomeningocele may be made clinically by the presence or absence of paralytic lesions in the legs.

The paralysis resulting from exposed neural tissue and damage to the spinal cord perhaps in utero and perhaps during birth does not bear the same close relationship to the level of the lesion that is found in traumatic paraplegia. It is difficult, therefore, to anticipate the degree of bladder paralysis or indeed of lower limb paralysis from estimation of the level of the lesion alone, though it is accepted (Sharrard 1962)that the foot deformities such as talipes equinovarus and the congenital dislocation of the hip found in myelomeningocele are paralytic lesions and not associated congenital abnormalities.

The mortality of myelomeningocele even with early closure of the back lesion, the use of antibiotics to control meningitis, and early ventriculocardiac shunt to control hydrocephalus, remains high. Of those children who die many are those with the more severe lesions. Nevertheless, many of the survivors remain severly handicapped in spite of multiple and often extensive operations. Some people believe that the more severe children should not be operated upon. Donald D. Matson (1968) states "when examination in the first day of life .... confirms total absence of neurologic function below the upper lumbar levels, custodial care only is recommended. It is recognised that some of these totally paralysed hydrocephalic patients recommended for custodial care survive for considerable periods of time. For these patients and their families, it is the doctor's and the community's responsibility to provide this care and to minimize suffering; but, at the same time, it is also their responsibility not to prolong such individual, familial, and community suffering unnecessarily, and not to carry out multiple procedures and prolonged, expensive, acute hospitalization in an infant whose chance for acceptable growth and development is negligible." These are the words of an experienced neurosurgeon who would not come to such a conclusion without a great deal of thought. He recognised that the condition is not necessarily lethal within a few months but he believes that these severe cases will die within a year or so. In fact 16% of them are alive after one

year (Lawrence 1966). 16% total survivors in an untreated series but some of these were minimal lesions. It is not clear how many of the severe untreated cases survive over a year but the care of a single completely paralysed child with unrelieved progressive hydrocephalus for months or years must make one think of the consequences of recommending "custodial care".

MATERIAL

Although the so-called pure meningocele may have associated problems of hydrocephalus with it, or may present later paralytic problems due to nerve root traction from the associated lipoma, the treatment of these children has not aroused the same heated arguments as have been aroused by the treatment of the open myelomeningocele with its more extensive disabling complications. An unselected series of 200 consecutive closures of open myelomeningoceles was therefore taken, 194 of them operated upon in the first 36 hours of life. These children were born between the middle of 1962 and the middle of 1964 and included all children admitted to The Children's Hospital, Sheffield, during that time, with open myelomeningocele except for two who were not operated upon because clinically they had sustained severe intracranial damage with probable ventricular haemorrhage and seemed unlikely to survive. These children were all operated upon by the same surgical team. There were 117 girls and 83 boys.

General Management

The aim in every case was early closure of the back lesion, operation being delayed only for the length of time required to have blood cross-matched for transfusion and for the operating theatre to be prepared. Post-operatively the babies were nursed in the neonatal surgical unit in incubators; antibiotic therapy was given in only those cases where there were specific indications such as early infection of the back lesion or the development of infection in the wound, the meninges, or the urinary tract. During the first admission baselines were established for the size of the cerebral ventricles and the state of the urinary tract. Clinical evidence of hydrocephalus such as an abnormally large head circumference at birth or the presence of separation of the lambdoid suture or evidence of abnormal increase in head circumference were indications for air ventriculography. Frequent bacteriological examinations of the urine were carried out and every child had an intravenous pyelogram before discharge; evidence of urinary tract infection or the presence of anatomical abnormalities in the urinary tract shown on intravenous pyelography were indications for more detailed urinary examinations such as micturating cystourethrography.

Routine outpatient examinations were carried out at the age of three months, six months, nine months, one year, and thereafter at six monthly intervals. Surgical intervention for the control of progressive hydrocephalus or for the achievement of adequate urinary drainage was undertaken if there were indications of its necessity at these routine visits. The orthopaedic problems which will not be dealt with in detail in this thesis were dealt with by orthopaedic colleagues.

Although a number of these children are now approaching the age of ten and the above pattern of treatment was established in Sheffield from 1960 onwards, it was felt that there would be insufficient children to review at ten years of age. In addition continued close personal contact has only been maintained with these children since the end of 1962. It was therefore decided to review the condition of the children at the age of five at a time when they were ready to start school.

An assessment was made on three counts with as much objectivity as possible. Firstly his intelligence quotient as measured by a psychologist was recorded; secondly his physical ability in locomotion as described by his mother or his teacher, rather than by the sometimes optimistic specialist who had treated him, and thirdly the degree of control of his urinary tract. On these three counts will the ability of the child to fit into society depend.

Classification

As previously mentioned the degree of paralysis of the lower limbs and pelvic floor in these children does not bear the same accurate relationship to the level of the lesion as is found in traumatic paraplegia; the damage to the spinal cord is much more variable than in a transverse section of the cord and indeed pathological abnormalities may be found in the spinal cord at entirely different levels from the level of the myelomeningocele. It has been the practice of our orthopaedic colleagues in all cases to make an accurate assessment of the innervation of the lower limbs at the time of admission of the child by estimating and recording both voluntary movement and response to faradiam. Such an assessment requires considerable skill and experience, especially in a baby who is colder than a normal child of the same age. A

LEVEL OF LESION

Level	Number	Per cent.	
T/L/S	38	19	
T/L	49	24.5	71
L/S	55	27.5	
L	27	13.5	
S	23	11.5	29
T	8	4	
TOTAL	200	100	

TABLE I

simple broad classification of the level of the lesion was therefore designed, one that could be easily estimated by any medical practitioner examining the child; 29% of the cases had a limited lesion involving only the lumbar, sacral or thoracic regions, the remaining 71% had more extensive lesions involving lumbo-sacral, thoraco-lumbar or thoraco-lumbar sacral regions (Table 1). Clearly these latter 71% patients would be expected to have more severe paralysis.

Deaths

Eighty-three of the children had died by the time they were five years old. This not inconsiderable mortality of 41.5% has been improved in more recent years. However, the mortality to a certain extent is selective since although the more severe lesions involving more than one region account for 71% of the cases they account for 84% of the deaths (Table 2, Fig. 2, Fig. 3). It is also notable that the curve of the number of children surviving drops most rapidly in the first three months for all levels of the lesion, but even after that time the curve is a steeper one in the more severe lesions.

The majority of those children dying in the early period died of intracranial haemhorrage or infection (Table 3). Ten children died of chest infections and these were mainly those who had very high lesions involving the dorsal spine, and consequently reducing respiratory excursions. Another 29 died of meningo-ventriculitis. Pneumonia remained quite a significant cause of death even in older children, again particularly in those with high lesions. Meningitis and ventriculitis occurring after the age of six months and causing death were usually the result of infections of the shunt systems for the hydrocephalus. As one would expect the renal tract played an increasing part in producing mortality in older children.

It is difficult to compare the mortality rate in this series with the mortality rate in similar children treated conservatively because admission to a special hospital for closure of the back lesion will also involve specialised care in a neonatal surgical unit. It seems rather unlikely, however, that the surgical interference does much to improve the immediate survival rate of itself, on the contrary two of the patients in the series died

NUMBER OF DEATHS

	T/L/S	T/L	L/S	L	S	T	TOTAL
0 - 6/52	19	12	12	2	2	1	48
6/52 - 3/12	1	2	-	2	-	-	5
3/12 - 6/12	1	3	3	2	-	-	9
6/12 - 1 yr.	1	1	3	1	-	-	6
1 - 2 yrs.	2	5	-	1	1	-	9
2 - 3 yrs.	2	-	-	-	-	-	2
3 - 4 yrs.	1	-	-	1	-	-	2
4 - 5 yrs.	-	1	1	-	-	-	2
TOTAL	27	24	19	9	3	1	83

TABLE 2

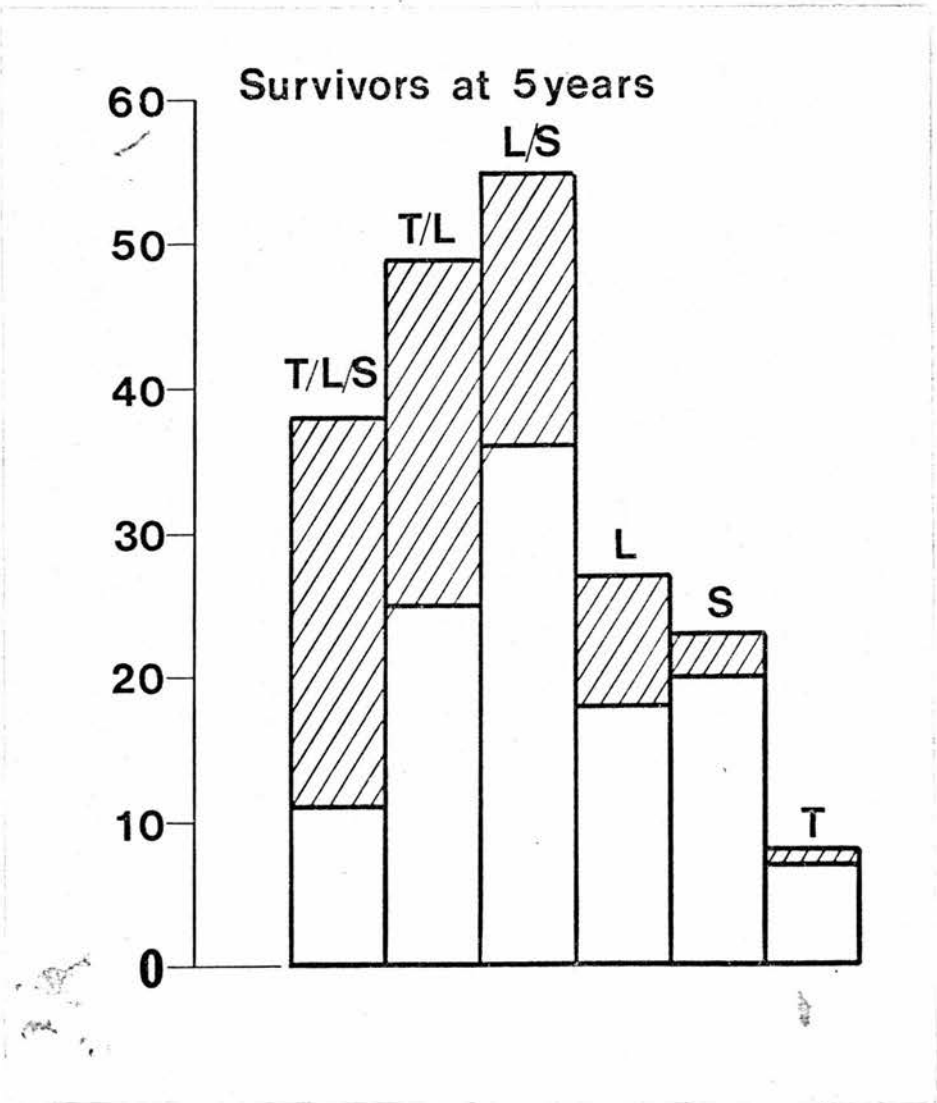

Figure 2

Cross hatching represents deaths

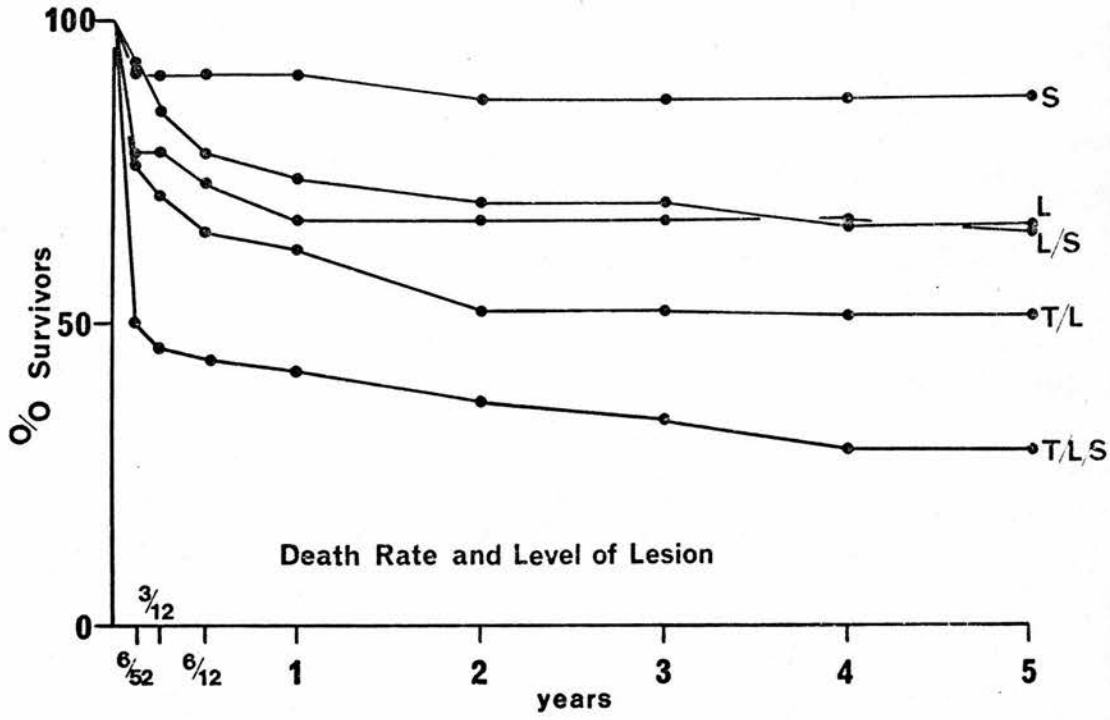

Figure 3 : Graph of survivors

AGE AND CAUSE OF DEATH

	6/52	3/12	6/12	1	2	3	4	5
i/c Haemorrhage	6	-	-	-	1(v)	-	-	-
Ventriculitis meningitis	29(2v)	2	2	2(v)	2(v)	-	-	-
Pneumonia	10	2	2	1	3	1	-	-
Renal	1	-	2	2	4	-	1	2
Valve malfunction	-	1	3	1	-	-	1	-
Operative	2	-	-	-	-	-	-	-
TOTAL	48	5	9	6	10	1	2	2

(v) indicates patients with valve

TABLE 3

as a result of the operation; both children had other abnormalities - one having a hypoplastic lung associated with his rib cage deformity and the other an intestinal obstruction of unknown origin occurring post operatively; yet both might have survived had the back not been operated on.

Long term survival however does appear to be affected. Sherrard, Zachary and Lorber (1967) showed that in similar cases 54% with closed backs survived to three years whilst only 32% of those with unclosed backs were alive at that time.

Seven children with less severe lesions who died under three months of age deserve more detailed consideration. Four of them died from ventriculitis; two from intracranial haemorrhage and one from gastro-enteritis. The two who had intracranial haemorrhages both had super added chest complications but they might be regarded as unavoidable deaths; their legs were good. Three others had good legs, the one who died of gastro-enteritis after discharge from the Children's Hospital and two of those who died of ventriculitis. It is these last two who cause particular concern; one had frank wound sepsis and the other had a well healed back wound but it could be argued that operative interference in each case contributed to the development of ventriculitis and subsequent death of the child.

HYDROCEPHALUS

Whilst it is true that the majority of children with myelomeningocele have associated hydrocephalus, not every child with hydrocephalus requires surgical interference. A number of systems of drainage of the cerebro-spinal fluid are available but none can be claimed to be curative because what they do is to control progressive hydrocephalus rather than to deal with the fundamental cause of hydrocephalus; they do this by short-circuiting the normal system of reabsorption of cerebro-spinal fluid. In the series under consideration the system used was direct drainage of the right lateral ventricle by a catheter inserted into that ventricle through a burr hole; the catheter is attached to a Holter valve which is bedded in the skull behind the right ear, and from the lower end of the valve a longer catheter is passed down into the neck into the internal jugular vein and this lower end is directed into the right atrium. Other efficient valves are available but at least during the time of the study no more reliable system had been discovered and we continued to use the one to which we were accustomed. The decision to establish ventriculo-cardiac drainage depended on evidence of increasing hydrocephalus first by the head circumference increasing at an abnormal rate and second by confirmation through air ventriculography. The air ventriculography was done by inserting a needle through the lateral angle of the anterior fontanelle into the ventricles withdrawing up to 10 ccs. of fluid and injecting a similar amount of air. X-rays were then taken in varying positions. Since ventriculo-cardiac drainage will relieve hydrocephalus of either the internal or the external variety, the air ventriculography was carried out more for getting an accurate estimate of the thickness of the cerebral mantle than for indicating the exact level of the obstruction to the flow of cerebro-spinal fluid. A cortical thickness of less than 15 mm. was taken as an indication for the insertion of a valve and a thickness of over 35 mm. was regarded as a degree of hydrocephalus not requiring treatment. Babies with measurements between these two were observed for a further period, some achieving an equilibrium and some continuing to grow and demanding valve treatment. (Lorber 1961).

One hundred and two of the total of 200 cases had a ventriculo-cardiac shunt established on the above indications. Whilst this figure represents only 51% of the total it must be remembered that of the 53 children who died within the first three months of life many would have required ventriculo-cardiac drainage should they have survived. It is also noteworthy that of the 142 children with more severe lesions 72 survived and only 20 of these survivors had no shunt, whereas in the 58 children with the less severe lesions, 45 survived of wh om 23 had no shunt. Clearly in this series the more severe the lesion the more likely it was that hydrocephalus would be bad enough to demand treatment. For example, of the 38 children with a thoraco-lumbo-sacral lesion 16 had a shunt, but of the remaining 22, 19 had died within the first three months of life and in fact 18 within the first six weeks; most of these children would have had shunts had they survived.

Once a shunt system has been established the patient is exposed to a new set of risks; he has an indwelling foreign body with the distal nd of it actually in the circulating blood, and he becomes dependent on the continuing efficiency of the shunt system. In the newborn child it is unusual for the child's life to be in danger from acute increase of the intracranial pressure due to the collection of increasing amounts of cerebro-spinal fluid; the sutures are open as is the anterior fontanelle, and the head is pliable enough to expand in response to the increasing pressure. When, however, the pressure has been kept at a more normal level, the head has grown and further ossification has taken place, interruption of the free flow of cerebro-spinal fluid through the shunt system will produce a rapid rise in intracranial pressure and an immediate danger to the child's life. Numerous procedures, therefore, are likely to be necessary to maintain the adequate function of the shunt system. Of the 103 valves inserted 61 cases required further operations and a total of 155 revision procedures were carried out on these 61 children up to the age of 5 years. A number required further operations later in life.

The reason for placing the distal catheter with its tip lying in the atrium is to ensure that the cerebro-spinal fluid is flowing into a fastly moving

current of blood and to discourage the clotting of blood around the tip of the catheter. Clearly as the child grows the relationship of the valve and its catheters to the original anatomical position must change and the tendency is for the valve to remain fixed over the mastoid process and for the distal catheter to rise relative to the atrium and lie in the superior vena cava or even in the internal jugular vein. Some workers believe that elective procedures should be carried out in order to reposition the valve and its catheters. (Tsingoglou and Forrest 1968). Pressure of work, experience that many shunt systems continue to function even when the valve is lying very high up in the neck, and experience as in one child in this series where valve revision was followed by temporary hemiparesis, has discouraged us from elective procedures of this nature. Only two of the children had elective revisions in this series; both had had the valve inserted under the age of eight weeks, in one case the lower catheter was lengthened at the age of 3, and in the other at the age of 4. Both these children, in fact, required later revisions because of blockage of the distal catheter.

Valve revisions

The indications for interference are shown in Table 4. At the first revision 36 children had blocked lower catheters and 10 had blocked upper catheters. Blockage of the upper catheter was usually brought about by clogging of the orifice in its tip with perhaps the choroid plexus, occasionally by change in the relative size of the ventricle causing the ventricular catheter to slip out of the ventricle altogether, occasionally by detachment of the ventricular catheter from the valve, the catheter being shown to be lying free in the ventricular cavity. Correction of these was by replacement of the ventricular catheter. Blockage of the lower catheter was primarily due to clotting around its tip and this usually occurred when the catheter had become relatively short for the child and lay in a vein rather than in the right atrium. Correction here would be in the first place by an attempt to lengthen the catheter through the same vein but this is difficult because the tip of the catheter lies in a little clot cavity lying in the side wall of the vein and it is rarely possible to push another new catheter further down; if this were not possible then an attempt would be made to pass the catheter

Indications for Revision

Revision number	1st	2nd	3rd	4th	5th	6th	7th	8th	9th	10th	
Blocked ventricular catheter	10	2	3	3	3	1	1	1			
Blocked venous catheter	36	11			1						
Blocked peritoneal catheter		10	13	4	1				1	1	
Both catheters blocked	8	2									
Infection	6	5	2	1							
Mechanical malfunction	1		1								
TOTAL	61	30	19	8	5	1	1	1	1	1	

Twenty seven additional procedures were required for shunt systems becoming infected following revision.

TABLE 4

into another vein in the neck, usually the external jugular, but again frequently, since this new catheter would end up in the same vein that had been used before, there was a tendency for it to obstruct. When the attempt to find a suitable vein in the neck had failed a drainage system was established, passing a long lower catheter subcutaneously over the anterior wall of the chest and inserting its distal end into the peritoneal cavity. Various attachments had been devised for the tip of this peritoneal catheter to discourage the formation of a sac of filmy membranous tissue around it which seems to be produced by the flow of the cerebro-spinal fluid, but none of these have been uniformly successful, and the frequency of blockage of the ventricular peritoneal system is shown by the indications for the later revisions (Table 4). After the lower end of the catheter in a vein in the neck has been blocked once or twice the majority of the distal catheters were placed in the peritoneum and therefore blocked peritoneal catheters formed an increasingly common source of trouble. After the third revision the numbers of blocked peritoneal catheters decrease again and this is because at that stage a further attempt has been made to find a vein in the neck when collateral circulation had opened up; this usually occurred when the child was perhaps 3 or 4 years old and following that the relative rate of increase of the distance between the lower end of the valve and the right atrium becomes much less and the children were established with a more satisfactory ventriculo-venous system once more. Table 4 shows one revision occurring from the sixth to the tenth time, these were in fact all in one patient.

Relation of degree of hydrocephalus to number of revisions

There was no significant difference in the number of upper end blocks in those children with larger ventricles; of the 61 children who had blocked valves 33 had a cerebral mantle under 17 mm. and 28 a mantle over 17 mm, and ten of each required revision of the upper end of the system on one or more occasions. If the number of systems requiring revision is related to the age at which the valve was inserted, however, (Table 5) about 62% of those inserted under 3 months of age required revision and only 48% of those inserted over three months. Of the 21 children who had shunts put

Valve Revision in relation to age at insertion

Age at insertion	Shunt	Shunts requiring revision	Percentage
Under 6 weeks	52	32	61.5
6 - 12 weeks	30	19	63.3
Over 3 months	21	10	47.6

TABLE 5

Infected Shunt Systems

Staph.Albus Valve Colonisation	3
Other infection in ventriculo-cardial shunt	7
Infection in ventriculo-peritoneal shunt	4
	14

TABLE 6

in at over 3 months of age 18 survived at 5 years and 5 of those 18 had an I.Q. of under 70. This is not very much different from the overall number of children with a low I.Q.; delayed shunt establishment might therefore be justified.

Infection of the shunt system

As shown in Table 4 six children required revision in the first instance because of infection of the shunt system. These and the eight who had later revisions because of infection have not had their further 27 operations included in Table 4, since infection raises its own particular problems (Table 6). Three children developed colonised valves, the colonisation being with staphylococcus albus: two were colonised after the first insertion, one after a revision. The explanation of this valve colonisation has never been very clear. It may be that it occurs as a result of infection at the time of insertion of the valve but it could also be due to colonising of the valve from incidental staphylococcus albus bacteraemia (Noble et al 1970). Holt (1970) showed in an experimental model that ascending infection could occur. In one of these children the shunt was removed and was never required again but in the other two after removal of the shunt there was a rapid increase in the intracranial pressure and this increase had to be controlled by external drainage with a tube passed into the ventricle and draining against gravity. In each case this tube passing from the ventricle to the exterior eventually led to further infection and the child died from ventriculitis.

One child died within four days of valve insertion of staphyloccus aureus septicaemia. In two cases the valve had to be removed within two weeks of its original insertion because of staphylococcus aureus septicaemia: one died the day the valve was removed and the other had a further shunt system inserted six weeks later which again became infected and was converted into a ventriculo-peritoneal shunt but the child died of a terminal intussusception associated with the peritoneal tube which had perforated and run from the stomach into the small bowel.

Four children developed staphylococcus aureus or pyocyaneus infections following revision of a ventriculo-cardiac system. In two after removal of the system and antibiotic therapy for four weeks a new system was inserted on the other side of the neck and the children remained under control: one required external

Ventriculo-Peritoneal Shunts

Made at 1st revision	19
Made at later revisions	10
Total	29

Results

No further revisions	5
One further revision	4
Two or more further revisions	7
Returned to vein after further revisions	7
Removed because of infection	2
Died from valve complication	4

TABLE 7

Valve Deaths

Unassociated with valve		
Pneumonia	2	
Renal	5	
Intracranial haemorrhage	1	
Mongol	1	
Fibrocystic	1	
Unknown	1	
		11
Associated with valve		
Over active valve	1	
Infection	7	
Pulmonary emboli	2	
Blocked shunt	3	
Gastroenteritis	4	
		17

TABLE 8

drainage for a period but survived with a left sided shunt and one died at home five months after establishment of a left shunt probably of chronic infection.

Four other children had ventriculo-peritoneal shunts which became infected; two died of fulminating meningitis; one survived after removal of the system, antibiotic therapy and insertion of a shunt on the other side; and the last, who had a dramatic faecal meningitis the lower end of the shunt being in the colon had her system removed and replaced, the new system infected and removed and eventually survived with no shunt system at all.

Taking the management of the 14 patients with infected shunt systems as a whole, one died before any procedure was carried out and another on the same day as the shunt system had been removed. Five had the shunt system removed and were treated energetically with antibiotics, one of these never required a shunt again and four had a left sided ventriculo-atrial shunt re-established. Of these four only one survived, two of the other dying later probably from complications other than their valves, and one of them died later from an infection of the new shunt.

Seven of the children having had the shunt system removed required continuing treatment to control the intracranial pressure, one had a Rickham reservoir inserted (this being a small silicone reservoir attached to a tube passed into the ventricle which can be aspirated rather than the ventricle itself) and another had a large burr hole made. These two children had their intracranial pressure kept at a lower level by repeated aspiration of C.S.F., either direct through the burr hole or through the reservoir. The other five children required external drainage systems, three of them before removal of the valve, the distal catheter merely being brought out through the skin to drain into a sealed container and two of them after removal of the valve, a tube being passed in through the skin into the lateral ventricle and draining against gravity. Of these children who required continuing control of their hydrocephalus only three survived, the one who had had the Rickham reservoir and two who had had exteriorisation of their valved shunt system with antibiotic therapy and later replacement of the valve system. It is notable that none of the children who had direct external drainage with no valve in the system survived.

One child who had repeated attacks of meningism without evidence of bacterial organisms but a high leukocyte count in the cerebro-spinal fluid was eventually shown to have a dermoid cyst discharging into the lateral ventricle; he presented as a repeated infection but is not included as a true infection.

Ventriculo-peritoneal shunts

In this series of cases the primary drainage procedure involved insertion of the distal catheter running from the Holter valve into the venous system. This is regarded as a procedure of choice and even when the original system had become occluded a farther effort was made to find another suitable vein for insertion of the distal catheter. Ventriculo-peritoneal shunt was therefore regarded as a less satisfactory alternative procedure and the results tend to confirm this impression.

Twenty-nine ventriculo-peritoneal shunts were established, 19 being made at the first revision, 10 at later revisions of the ventriculo-cardiac systems (Table 7). Five of these children required no further procedures and four required only one revision during the first five years of their lives. These could be regarded as fairly satisfactory results. The remaining 20, however, could not be regarded as satisfactory. Seven required two or more further revisions and another seven reverted to a venous shunt after multiple revisions. Two had the system removed because of infection, one of these eventually had a venous shunt re-established and the other had no further shunt system inserted. Seven of these children died, two from renal complications in whom the shunt could be regarded as playing little part, one from renal complications but post mortem also showed multiple pulmonary emboli which might have been attributed to the previous venous shunt which had been infected, and four as a direct result of ventriculo-peritoneal shunt complications, three of meningitis and one from raised intracaranial pressure when there was a block of the shunt system.

Deaths

Eleven of the children with valves died, probably of causes unrelated to their hydrocephalus (Table 8), two died of pneumonia, five died of renal causes and

and one probably of an intracranial haemorrhage, post mortems showing that the hydrocephalus was controlled. Three died at home, and had no autopsy: one was a Mongol and one had fibrocystic disease and both these children had had repeated chest infections; the third died two days after his discharge home, and only twelve days after the insertion of his valve. His head circumference had come down, he had no infection and it seemed likely that he died of inhalation of vomit.

Seventeen children died from causes which could be regarded as definitely associated with the valve. One died apparently from an overactive valve with too rapid decompression. Seven died early after infection and these have already been discussed. Two died up to five months after having been shown to have an infected valve system and septicaemia and both were shown to have multiple pulmonary emboli. Although the emboli had been regarded as complications of ventriculo-atrial drainage it is unlikely that they can be regarded as the primary cause of death and they are much more likely to be incidental findings in children who have died from other causes (Emery 1971). Possibly in these two children the cause of death was again septicaemia. Three children died as a result of blockage of the shunt system, two of them being acute deaths about four months after the insertion of their valves, death being sudden and in each case in children under six months of age; the third death due to block occurred in a child who died on the operating table at the time of revision of a ventriculo-peritoneal shunt.

Four children died with evidence of gastro-enteritis. These are an interesting group of patients, two of them died within the first three months of life, one at the age of one year and one at the age of three. The three babies certainly all had gastro-enteritis with severe dehydration and were all known to have a functioning shunt. The older child who also had severe gastro-enteritis was shown to have a functioning shunt and considerable coning through the foramen magnum at post mortem examination. It seems possible that these children may all have had an upset of the dynamics of the cerebro-spinal fluid circulation due to the dehydrating effect of the gastro-enteritis, and there is no doubt that other children who did not die have been shown to have an apparent failure of the shunt system when

suffering from intercurrent pyrexial illnesses.

URINARY TRACT

For its insidious onset and progress, disease of the urinary tract is notorious and in children the danger of extensive destruction of renal tissue occurring without clinical evidence of disease is even greater than it is in adults. General management of the child with regard to the urinary tract has therefore been directed at the early recognition of deviations from the normal and the institution of appropriate measures to correct those deviations.

Although the incidence of anatomical abnormalities in the urinary tract is higher in children with myelomeningocele than in the normal population, (Roberts 1961, Emery 1969) these abnormalities are of far less clinical significance than the abnormalities which are secondary to the neuropathic bladder. Basically almost every child with an open myelomeningocele will have some degree of abnormality of bladder innervation, though in the small localised lesions this interference may be minimal. The ultimate survival of the child will depend on the efficiency with which he can dispose of the urine secreted by his kidneys. If the bladder dysfunction is of the type which is conducive to the maintenance of an appreciable amount of residual urine in the bladder then that urine is likely to become infected and a combination of infection and back pressure will lead to progressive dilatation of the upper urinary tract. Vesico-ureteric reflux plays some part in this though it is not always present in those cases in which the kidneys show the effects of back pressure.

Bearing these factors in mind some authors (Smith 1965) believe that in all cases an early diversion of the urinary stream from the bladder should be undertaken. Since the results in the series of cases described here show that at the age of five 24 out of the 117 survivors had almost normal urinary control, such blanket recommendations for operation seem to be unjustified, and the principle in our series was that each child should be carefully watched for early evidence of deviation from normal and that surgical interference should be undertaken only when there was a positive indication for it.

The routine management involved first the primary assessment of the child. Although we have not been able to make such an accurate correlation between the

level of the lesion and the probably neurological pattern of the bladder as was done by Stark (1968)it is certainly true that if the primary examination shows normal function in either one or both lower limbs it is unlikely that there will be serious bladder dysfunction, and if the anus is patulous it is certain that there will be some degree of bladder incontinence. More important perhaps is the information usually gained by the nursing staff in the first few days of life; they are instructed to attempt suprapubic expression of the bladder in all these children. If suprapubic expression is possible then it can be assumed that there is abnormal innervation of the bladder, and as a routine each time the baby is handled the bladder will be emptied by suprapubic expression. If suprapubic expression of the bladder is not possible then either the bladder innervation is normal or there is sufficient outflow obstruction produced at the bladder neck or at the external sphincter to prevent even manual expression of a full bladder; further information can be obtained from watching the child and from seeing a good stream of urine passed either spontaneously or as a reflex to a cold hand or slight pressure on the lower abdomen. Bimanuel palpatation of the bladder with a finger in the rectum gives useful information with reference to the amount of urine in the bladder. Specimens of urine will be taken regularly for bacteriological examination and frank infection will immediately alert the clinician to the likelihood of some interference with normal flow of urine.

Radiological investigations include an intravenous pyelogram in every case in the first few weeks of life to establish the normality of the upper urinary tract. Frank hydronephrosis may be shown but this investigation may again alert the clinician to further investigations, since a dilated lower ureter or perhaps a trabeculated bladder may give early warning of outlet obstruction. Thus, abnormality in the intravenous pyelogram suggesting dilatation of the upper urinary tract, frank urinary infections; or the repeated finding of an appreciable amount of residual urine are taken as indications for further investigations. The most useful routine investigation here is the micturating cystourethrogram performed by passing a catheter up the urethra and injecting opaque medium into the bladder. This examination will indicate the presence of vesico-ureteric reflux, the degree of trabeculation of the bladder wall and the character of the urethral flow. Whilst normally called the micturating cystourethrogram in practical terms it is much

Urinary Procedures

Bladder Outlet Improvement	
Pudendal neurectomy	1
Y.V. Plasty	7
Transurethral resection	2
Sphincterotomy	2
Ureterostomy	8
Bowel conduit	
Ileal	22
Sigmoid colon	3
Revision procedures	10
	55

55 procedures in 34 patients

(3 more patients awaiting diversion)

TABLE 9

Upper Urinary Tract following Surgical Procedures

Normal I.V.P. at 5 years	10
Controlled hydronephrosis at 5 years	16
Progressive renal destruction	4
Dead	4
	34

TABLE 10

more commonly a voiding cystogram, the investigator depending on suprapubic pressure to fill the posterior urethra. An additional value of the examination is that it will give visual evidence of residual urine in the bladder after expression.

Residual urine could be regarded as the key to the maintenance of the integrity of the upper urinary tract. Treatment is designed to avoid residual urine and its stagnation. This may be achieved by regular suprapubic expression alone and in appropriate cases a routine is established from the earliest days of the child's life, first by the nurses and later by his mother, of urinary expression each time the baby is handled. In some cases where cystourethragraphy has demonstrated narrowing of the bladder outlet then that outlet has to be enlarged. If widening of the bladder outlet does not allow free drainage of urine then a supravesical urinary diversion is required, either by direct cutaneous ureterostomy or by the use of a bowel conduit.

In this series 34 of the patients underwent surgical procedures on the urinary tract during the first five years of their life and three more at the age of five were awaiting urinary diversion. (Table 9)

Four of the children who had had operative procedures carried out on the urinary tract died, all of advancing renal disease. (Table 10) One died at seven months of age, a second under two years, and the third and the fourth in the fourth year of their lives. Of the survivors ten had a normal upper urinary tract shown on the intravenous pyelogram at the age of five, 16 had hydronephrosis which was not advancing and appeared to have been controlled by the urinary procedures, and four had severe hydronephrosis and upper urinary tracts which were probably deteriorating. The figures for the state of the urinary tract at the age of five in the series as a whole are poor but these figures in children with urinary tracts that required surgical intervention give rise to particular concern. It is worth considering the individual surgical procedures even though this series is small.

(a) Pudendal neurectomy

Pudendal neurectomy is an established procedure in the management of the neuropathic bladder, the rationale being that the pudendal nerve supplies the external sphincter muscle and if this nerve is divided on one or both sides the

balance between the detrusor activity and the sphincteric spasm will be altered and emptying of the bladder will be easier. Theoretically the operation would appear sound but its success depends on two things, first that the contracting external sphincter is the obstructing agent and second that the detrusor muscle of the bladder is sufficiently strong to extrude urine once the sphincter mechanism has been weakened. In our experience this has not been a successful operation in children with myelomeningocele. It was carried out in only one child in this series who first had a pudendal neurectomy, later had a Y.V. plasty of his bladder neck because of failure of improvement of bladder drainage, whose upper urinary tract continued to deteriorate necessitating a urinary diversion through an ileal conduit and who in fact continued to deteriorate further partly as a result of failure of the conduit on one side, and at the age of 5 had had one nephrectomy and had severe hydronephrosis of his sole remaining kidney. This child incidentally after the age of 5 had an incident of unconsciousness associated with malignant hypertension.

(b) Y.V. Plasty

Seven children had plastic procedures on the bladder neck, a Y shaped incision being made over the lower anterior wall of the bladder extending on to the posterior urethra which was closed as a V thus widening the bladder outlet; in most children also a wedge of tissue was excised from the posterior aspect of the bladder outlet. Only one of these children retained a satisfactory upper urinary tract after this procedure: one had an additional transurethral resection of the bladder neck and external sphincter before the age of 5 and had a static hydronephrosis at the age of 5 though later he had a urinary diversion. One had a later urinary diversion through a bowel conduit and had a satisfactory upper urinary tract at the age of 5. Two had several more procedures with urinary diversions and still had progressive hydronephrosis at the age of 5, and two died, one after progressing to a urinary diversion and one having only had a Y.V. Plasty.

(c) Cutaneous uretorostomy

A cutaneous ureterostomy can only be made successfully when the ureter is dilated, the normal sized ureter being too narrow to make an effective spout on

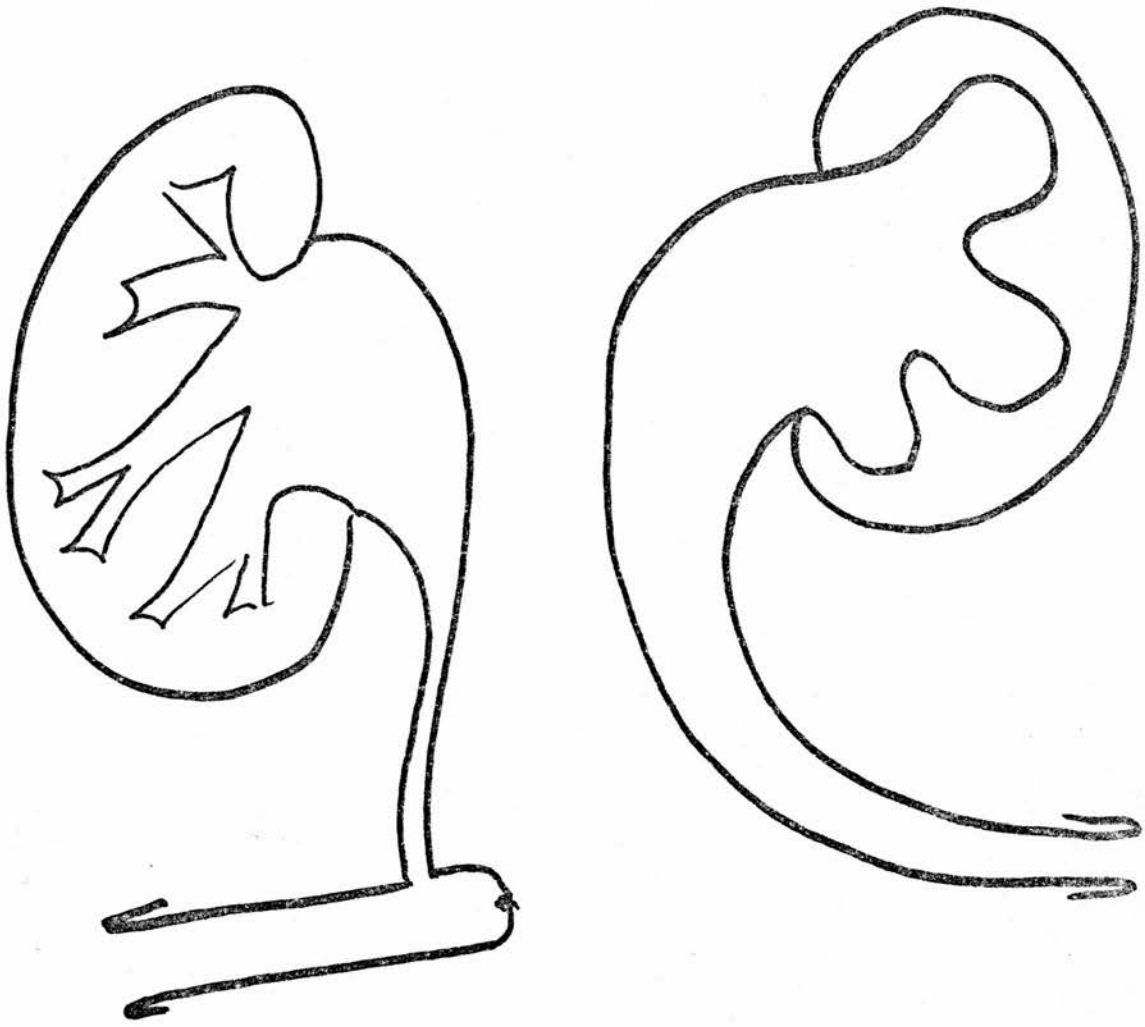

Figure 4 : Diagram of ureterostomy
and ileal loop

the abdominal wall and indeed often being too short to reach the surface. Ureterostomy therefore can only be regarded as a salvage procedure in attempting to achieve drainage of a urinary tract which has already become dilated as a result of obstruction and possibly associated infection. At the same time the dilated ureter can form a very satisfactory spout brought out through the abdominal wall at the position of choice for the suitable application of a collecting apparatus, and once the ureter has become moderately dilated there seems little advantage in using an isolated loop of bowel as a conduit; thus a few of the children who had ureterostomies as salvage operations may be expected to retain these ureterostomies as a permanent supravesical diversion. (Lister et al 1968).

Of the eight children who had cutaneous ureterostomies one was a child with a solitary kidney who had early hydronephrosis and a slight hydroureter and had the ureter brought out as a terminal ureterostomy; there was a tendency to stenosis and an additional pelvi-ureteric obstruction and the child was awaiting the formation of an ileal conduit at his fifth birthday.

In two children a supravesical urinary diversion was indicated because of persistent residual urine, frequent urinary infections, and evidence of upper urinary tract dilatation. However, the upper urinary tract dilatation affected the left side only, the right side being normal, and it was deemed inadvisable to drain the good right kidney and the bad left kidney into the same ileal conduit since this might encourage infection of the good kidney; indeed it was questionable whether the left kidney was worth preserving since its function was unknown. A ureterostomy was made on the left side, therefore, and the right side was drained through an ileal loop. (Fig. 4) In one case the left ureterostomy was later moved from the skin and attached to the ileal loop but at the age of five an intravenous pyelogram showed non-function on the left though the right kidney was working well; in the other case the child was left with a ureterostomy on one side and an ileal loop on the other side and both kidneys showed improvement in the intravenous pyelogram at the age of five.

Three children had loop ureterostomies made as emergency procedures when the child was in serious renal deficit. One of them who was only two months old was found at operation to have such a friable right ureter that it was avulsed

at the pelvi-ureteric junction and on the right side the kidney was drained using the appendix as a conduit; the left side had a direct cutaneous loop ureterostomy, but the child died of renal failure before he was six months old. Another child died of renal failure at the age of fourteen months after bilateral ureterostomies. The third child had bilateral ureterostomies made when he was eighteen months old and in considerable uraemia; he survived but at the age of 5 had progressive hydronephrosis. Two children had terminal ureterostomies done as emergency procedures, both of them eventually required a revision to a bowel conduit drainage, and one had continuing severe hydronephrosis at the age of 5, and the other had had one kidney removed for stones and progressive hydronephrosis in the other kidney, at the age of five.

Bilateral ureterostomies, therefore, carried out as salvage procedures, whilst likely to save the life of the child temporarily have not allowed very much in the way of recovery of the upper urinary tract. Those where one side only has been affected preoperatively have been more successful.

(d) Bowel Conduits

Bowel conduits made for drainage of the urine have been widely used for the diversion of urine from a malfunctioning bladder. (Cook, Lister 1968). Loop diversions, however, do not invariably control advancing urinary disease (Pekarovik et al 1968) and occasionally present problems of electrolyte reabsorption (Cook, Franks et al 1968). It has not been our policy, therefore, to divert the urine as a routine in all cases showing bladder malfunction. Operation has been carried out on two grounds, first, early evidence of deterioration of the upper urinary tract and second, a social diversion when a girl is unable to remain dry for an acceptable period and can be made more comfortable by wearing an appropriate appliance for collecting urine draining from a spout on the abdominal wall. Incontinent boys with a normal upper urinary tract can be successfully managed by wearing a penile appliance.

Twenty-five children had bowel conduit diversions, in 22 the ileum was used and in three the colon. At the time of this series it was thought that the

choice of sigmoid colon or terminal ileum for the formation of a conduit was mianly a matter that involved the individual choice of the surgeon or possibly the anatomical arrangement in an individual child; in fact operation was done through a transverse suprapubic incision which allowed the use of either ileum or sigmoid colon without altering or extending the incision. Later investigations in our research unit indicated that pressure within the sigmoid loop is often high and there may be further trouble for children with this type of loop, (Cudmore 1971) with continuing dilation of the upper urinary tract. In fact, in the three children in this series who had sigmoid loops made the indication in each case was a social one and the intravenous pyelogram at the age of five showed a normal upper urinary tract, except in one child who had had a nephrectomy carried out because of a technical error at the time of the operation; her remaining kidney, however, had hypertrophied and was functioning well.

Of those who had ileal conduits made, eight girls had the procedure carried out for social reasons and all these children had a normal intravenous pyelogram at the age of five. Three were children who had previously had bladder neck procedures carried out, one died of renal failure, one had had one nephrectomy and persistent hydronephrosis in his remaining kidney at the age of five, the third had an intravenous pyelogram that showed some improvement in the hydronephrosis. Five were children who had previously had ureterostomies done and of these only two showed some improvement of their renal function, the remainder had uncontrolled hydronephrosis or had already lost a kidney at the age of five.

Six children had ileal loops made because of evidence of increasing upper urinary tract dilatation. At five years two showed static hydronephrosis with no deterioration, two showed continuing increased hydronephrosis and two showed severe dilatation with stones. Thus again the results from supravesical urinary diversion by means of using a bowel conduit only rarely show improvement of hydronephrosis that was present at the time of the operation and only in those cases where the upper urinary tract was anatomically normal before operation could one expect to maintain this satisfactory state.

ORTHOPOEDIC PROCEDURES

Much has been written about the management of the paralytic deformities in myelomeningocele (Sharrard 1964, Sharrard et al 1968). A detailed analysis of the orthopaedic procedures required in this series of cases has not been carried out, partly because a considerable number of the children were operated on in other hospitals than the Children's Hospital in Sheffield and partly because it was not intended that the orthopaedic side should be covered in this consideration. However, it is of some interest that in the Sheffield Children's Hospital alone 332 orthopaedic procedures were carried out on these children before the age of five; it must be borne in mind that the majority of those children who died had died before any orthopaedic procedures could be undertaken on them. In addition to the essential stabilisation of the hip, correction of foot deformities, and appropriate tendon transplants to make full use of such muscles as are available, the orthopaedic surgeon is also concerned in the rather common lower limb fractures particularly fracture of the femur (James 1970). The severe kypho-scoliosis associated with myelomeningocele in some cases, both in the unusual lesion when only half of the cord is exposed (Duckworth et al 1968) and in the severe lumbar lesion where there is an acute kyphos of the lumbar spine, may also require orthopaedic treatment. Particularly in the kyphosis, osteotomy has been carried out in the neonatal period in some cases to help in the closure of the back at primary operation.

THE COST OF TREATMENT

Table 11 shows the estimated cost of treatment for this series of patients, set out as a balance sheet. The average cost of maintenance of an in-patient bed in the Children's Hospital in Sheffield over the seven years from 1962 - 1969 was £70 per week and out-patient attendances had been costed at £2.40. Ambulance and social services as well as appliances can be approximately estimated but the cost of treatment in children in other hospitals which a number of children received, the cost of general practitioner services for these children, local authority contributions to their care and the personal costs to the family of maintenance of a handicapped child have not been included. Heavy burdens other than financial ones are imposed on the families of handicapped children and the cost of such things as a broken home cannot be estimated in pounds and pence.

The total figure of £2053 per survivor is quite surprisingly low and even if the total cost were set against the number of normal or near-normal survivors the figure becomes £4000 each. The cost of maintenance of these children of course does not stop at the age of five. Fifty-three of the 117 survivors went to ordinary schools but for the remainder schooling would be costing about half as much again, in special establishments. Nor was treatment complete in all cases at the age of five, further orthopaedic and renal procedures being required in many cases in addition to further revisions of the shunt systems.

BALANCE SHEET

DEBIT 200 BACKS CLOSED		CREDIT 117 SURVIVORS AT 5 YEARS	
3017 Inpatient weeks	£217,224	Normal or near normal children	60
3000(estimated) O.P. visits	6,000	Moderately handicapped	23
Ambulances, social services	9,000	Severely handicapped	28
Appliances	8,000	Very severely handicapped	6
	£240,224		117

COST PER SURVIVOR £2053

Ordinary schooling for 50. Special school for 60.

Table 11

FIVE YEAR ASSESSMENT

Each survivor except three was seen at the time of his fifth birthday; of the three who were not seen, in two a written medical report was obtained indicating their progress and the third a child who had had a thoracic lesion with minimal nerve involvement was believed to be well.

Intelligence Quotient

A research psychologist measured I.Q's using the Stamford Binet test Form L.M. In some children extra tests were added e.g. Williams test for those with defective vision and Raven's progressive coloured matrices for those with severe motor defects.

Table 12 shows the intelligence quotient of the survivors. They were divided into those who had an I.Q. of over 80, those from 60 - 80 and those under 60, these being accepted as normal, educationally subnormal and entirely subnormal, from the point of view of schooling. Eighty of the children fitted into the normal category; a few had an outstandingly high I.Q. but the majority tended to be in the lower limits of normality. Thirty-six were estimated as being educationally subnormal and this has been taken to indicate a considerable handicap when associated with other physical handicaps; nevertheless it must be remembered that a number of men with an I.Q. of 70 are able to hold down a job and in fact support a family. It might also be said that in the presence of other severe handicap a low I.Q. may be a positive advantage to the individual. One child was severely subnormal.

If the three more extensive and the three less extensive lesions are grouped together it is evident that 29 (40%) of the 72 more severe lesions had a less than normal I.Q. whereas only eight (18%) of the 45 less severe lesions were in this category.

Micturition

Although lack of control of the bowel sphincters is clearly a disability on the whole it is a manageable one; it was found to be rare for mothers to have difficulty in bowel management or at least to complain of it. The children tend to be rather constipated and a bowel evacuation is usually obtained once or twice a day in response to suprapubic pressure perhaps with additional perineal pressure or a digital evacuation. Urinary incontinence, however, presents a much greater problem and apart altogether from difficulties with ulceration and

Intelligence Quotient of Survivors

	Normal		E.S.N.		S.N.		Total	
T/L/S	6		5		-		11	
T/L	12	43	13	28	-	1	25	72
L/S	25		10		1		36	
L	12		6		-		18	
S	18	37	2	8	-	-	20	45
T	7		-		-		7	
TOTAL	80		36		1		117	

TABLE 12

Micturition in Survivors

	Near Normal		Acceptably Dry		Diversion		Wet		Total	
T/L/S	-	5	8 (3)	35	1	21	2	11	11	72
T/L	1		8 (1)		9		7		25	
L/S	4		19 (5)		11		2		36	
L	5	19	8	16	2	5	3	5	18	45
S	8		7 (1)		3		2		20	
T	6		1		-		-		7	
TOTAL	24		51		26		16		117	

Bracketed figures indicate boys wearing penile appliance

TABLE 13

the anaesthetic skin of the perineum and thighs, the constant wetness creates considerable problems once the child is past the normal nappy age.

In this series 24 children had normal or near normal micturition and could be expected to be able to take their normal place in society as far as this was concerned. This is a rather higher figure than might be expected. It includes most of those children with a hemimyelocele and a lot of the simple lesions especially thoracic ones. One child with an extensive thoraco-lumbar lesion rather surprising fell into this category but serves as a reminder of the variable histology and the unpredictable end results. Another 51 were regarded as being acceptably dry. This implied in boys the successful wearing of a penile urinal or in girls or boys the ability to remain dry for at least two hours at a stretch, usually following bladder expression; these children again were regarded as individuals who could live in normal society without embarrassment.

Table 13 again shows within the more severe group of lesions only 40 (55%) out of 72 had reached this acceptable situation, whereas in the less severe lesions 35 (77%) out of 45 had achieved it.

Locomotion

With regard to locomotion 32 children were walking well without appliances, 67 required calipers but could walk, 18 were mainly confined to a wheelchair. This last group is perhaps an important one because although a number of these children were able to move with the help of calipers and sticks, those who cared for them, particularly in special schools, found it more convenient and quicker to move them around in a wheelchair; it may be that some of those who were mobile at the age of five with calipers would find as they grew older that a wheelchair was more convenient. However, a child walking with or without calipers was regarded as being a person who could live in normal society, whereas one who was confined to a wheelchair was regarded as having a considerable handicap.

Once more looking at the two groups of major lesions and minor ones we find that 17 out of the 72 (23.6%) of the major lesions ended up chairbound, but only 2.2%(1 out of 45) of the minor lesions. (Table 14)

Locomotion at five years

	Walking well		Calipers		Mainly chair		Total	
T/L/S	-	7	7	48	4	17	11	72
T/L	1		17		7		25	
L/S	6		24		6		36	
L	3	25	14	19	1	1	18	45
S	16		4		-		20	
T	6		1		-		7	
TOTAL	32		67		18		117	

TABLE 14

Table 15 is an attempt to take all these disabilities together. Sixty children had an acceptable brain, bladder and legs, 17 of them being perfectly normal; 23 were mildly handicapped with only one system being at fault; 28 were severely handicapped two systems causing trouble and 6 who had all three systems in trouble were regarded as very severely handicapped children.

Obviously those with a primary lesion extending into two or more areas of the spinal column in general are the most severley handicapped (Table 15). This can lead to the rather facile conclusion that the worst cases end up with the worst disabilities. It is, however, not true that all the bad cases in the first place are going to do badly because five thoraco-lumbo-sacral lesions, six thoraco-lumbar lesions and 17 lumbo-sacral lesions all ended up as near normal individuals, 11 of them being almost completely normal.

Future of the survivors

Figure 5 shows these end results as segments of a circle. 41.5% of the patients are dead at the age of five and 30% are normal or near normal. This leaves 28.5% who will depend on the community in varying degrees for their support during the rest of their lives. Some of them may be expected to be able to earn a living, but it would be unrealistic to presume that any of those with a subnormal I.Q., whether they also had bladder or lower limb involvement or not, would be likely to be independent members of society. There were 37 such children and they thus represent 18.5% of all cases of open myelomeningocele, or 31.6% of the survivors.

Again it would be wrong to expect all these 37 children to survive to normal adult life. Of the children with less than an average I.Q. 32 had ventriculo-atrial shunts to control their hydrocephalus and 24 of these had already had revisions (Table 16); it would seem likely that more revisions would still be required in these children and the child's life could be at stake at any time the system was blocked.

Perhaps more important from the point of view of long term survival is the state of the upper renal tract in the survivors. Table 17 shows the state of

Quality of Survival

	Dry Walking I.Q.> 80	Dry Walking E.S.N.	Dry Chair I.Q.> 80	Diversion or wet Walking I.Q.> 80	Dry Chairbound E.S.N.	Diversion or wet Walking E.S.N.	Diversion or wet Chair I.Q.> 80	Diversion or wet Chair E.S.N.
T/L/S	5	-	-	1	3	1	-	1
T/L	6	1	-	4	2	7	2	3
L/S	17 (2)	2	1	6	3	5	1	1
L	11 (3)	2	-	1	-	3	-	1
S	14 (8)	1	-	4	-	1	-	-
T	7 (4)	-	-	-	-	-	-	-
TOTAL	60 (17)	6	1	16	8	17	3	6

Figures in brackets indicate completely normal

TABLE 15

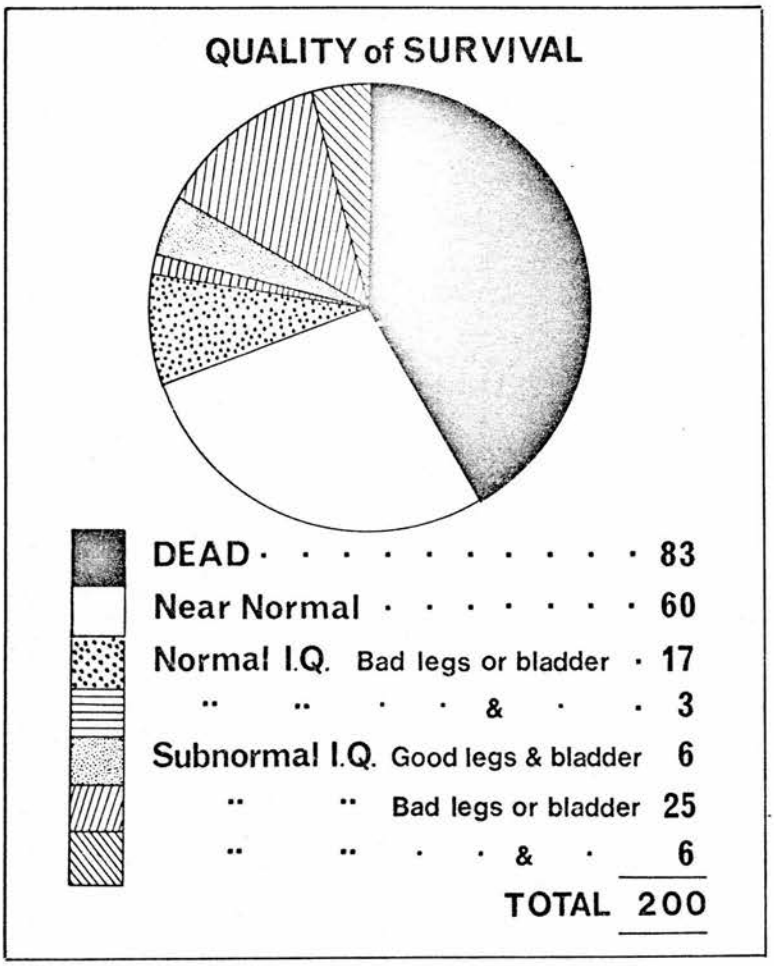
QUALITY of SURVIVAL
DEAD 83
Near Normal 60
Normal I.Q. Bad legs or bladder . 17
" " . . & . . 3
Subnormal I.Q. Good legs & bladder 6
" " Bad legs or bladder 25
" " . . & . 6
TOTAL 200

Figure 5

Shunt and I.Q. (Survivors at five)

	Normal I.Q.	E.S.N.	Total
No Shunt	38	5	43
Shunt (no revision)	13	8	21
Shunt and revision	29	24	53
Total	80	37	117

TABLE 16

Upper Renal Tract in Survivors

	Normal	Controlled	Deteriorating	Total
T/L/S	7	2	2	11
T/L	14	7	4	25
L/S	27	8	1	36
L	16	2	-	18
S	19	1	-	-
T	7	-	-	7
TOTAL	90	20	7	117

TABLE 17

the upper renal tract in the survivors as distinct from the function of their bladders. In 90 the upper renal tract remained normal, in 20 dilatation and deterioration appeared to have been controlled by surgical procedures, but in 7 the hydronephrosis which was already present was increasing and was not controlled by urinary diversion or other surgical procedures; the kidneys in these children are subject to persistent back pressure from obstruction to the urinary outflow and/or repeated damage from pyelonephritis with consequent reduction in functioning tissue and renal reserve. In fact already since the end of the five year follow-up two of these children have died and it is likely that some of the older ones will have the problems of hypertension added to their other complications. However, even if all those recognised as having deteriorated upper renal tracts were to die and half of the remainder we would still be left with at least 10% of the original cases growing to adult life as seriously handicapped persons.

ASSOCIATED ABNORMALITIES

In a series of this size it is not surprising that a number of the rare variations of a lesion would appear. Six children had the myelomeningocele affecting only one side of the cord, a lesion which has been called hemimyelocele (Duckworth et al 1968). These children present a special problem; they usually have completely asymmetrical paralysis, one half of the cord only being affected and thus one leg being perfectly normal. Provided there is full sacral innervation on one side at least, sphincter control is likely to be normal and the upper urinary tract is likely to preserve its normal function also. However, the patients do tend to have very severe scoliosis often with hemivertebrae and in addition many of them have a bony spur passing between the two sections of the split cord which may produce spastic paralysis due to traction on nerve roots. One of the children with a hemimyelocele also had a lipoma of the cord affecting the opposite side of the split cord; lipoma of the cauda equina is another variant of spina bifida but is not usually associated with open myelomeningocele (Lorber et al 1965, Emery et al 1969).

Nine children had renal abnormalities which were primary and not due to the effects of the myelomeningocele. Such abnormalities have been shown by others to be commoner in children with myelomeningocele (Roberts 1961, Emery et al 1964). There were three horseshoe kidneys, five solitary kidneys and one which had a complete duplex system.

Four children had alimentary abnormalities one with an anal stenosis, and one with an imperforate anus; one child had a duplication of the ileum and one child had pyloric stenosis. Two children had severe chest deformities, one in fact died at the primary operation of an associated lung hypoplasia, another child with bone abnormalities was one who had micrognathia and the Pierre Robin syndrome. There were two other associated abnormalities, one Mongol and one child with fibrocystic disease.

POTENTIAL DEVELOPMENTS

Although most of the operations performed on the children discussed in this thesis were designed to prevent progression of disease rather than to save life, there is no doubt that the survival rate at the age of five is very much higher than it would be in an untreated series of similar cases; operations designed to prevent progression of disease are likely at least to prolong life. The five year survival rate of 58.5% is low but not the first area in which to seek improvement. The quality of survival is far short of what might be desired. If one were looking for improvement in the end results one must either alter and improve the treatment given to the cases, or one must alter the nature of cases admitted to the series. Discussion will therefore review firstly those points at which treatment might be altered and improved, and secondly the possibility of concentrating effort on those patients in whom it can be expected that treatment will be rewarded with a successful result, and excluding from treatment those cases in which the prognosis is very poor.

Closure of the back lesion

It is a basic surgical principle that open wounds should be closed within as short a time as possible and it has been shown in myelomeningocele that considerable advantages are to be gained from early closure of the back lesion (Sharrard et al 1963). It seems quite clear that further damage to exposed neural tissue is avoided by covering it with dura mater, fascia and skin, and might be said that this primary closure was indicated on such basic surgical principles, that there was no need to show evidence of success but rather that if a primary closure were not to be performed then evidence would have to be shown that the procedure actually did harm.

Twenty-nine children who died of meningo-ventriculitis in the first six weeks of their lives may be of some importance for consideration here. Certainly five of them were shown at post-mortem to have upper urinary abnormalities, four already had hydronephrosis. Two had ventriculo-cardiac shunts established and died shortly thereafter due to infection of the shunt system. In the remaining 22 ventriculitis was the sole cause of death; eighteen were

extensive lesions; any of which could well have vecome infected even had they not been operated on, merely because of their extent. Four, however, were children with minor lesions, two affecting the sacral area only and two affecting the lumbar area only and all these were likely to have had limited paralysis and good function, had they survived. Two of these children in fact had good legs at the first assessment, as did two more who died at six to twelve weeks of age of ventriculitis. These patients developed their ventriculitis as a result of wound breakdown and infection and might have survived had they not been operated on. In the child who already has complete paralysis at birth with a wide separation of the skin edges, often accompanied by a severe kyphosis, then closure of the back lesion is almost certain to break down and it is likely that infection of a broken down wound is worse than infection of the unclosed lesion. From the point of view of survival of the child, therefore, a case can be made for not attempting to close the very wide extensive lesion where there is already complete paralysis of the lower limbs: a controlled trial of closure or non-closure in such cases has been considered. In the lesser lesion, however, the risk of loss of such function as is present at birth due to further damage to the neural plaque is probably greater than the risk of infection resulting from surgical interference; early primary closure should therefore be carried out, but recognising the incidence of infection following this procedure meticulous attention must be paid to asepsis before and during operation.

Another aspect in the prevention of infection has been considered though not analysed in this series of cases. Experience has shown that the broken down back wound may heal more quickly once the leak of cerebro-spinal fluid has stopped. During a period of six months from July to December 1967 eighteen children had ventricular drainage systems established during the first week of life deliberately to reduce leakage from back wounds; six of these children died, four with ventriculitis and septicaemia. A controlled trial was started to elucidate the effect of this early reduction of cerebro-spinal fluid pressure on the healing of the back wound. A series of 56 children were studied,but the problems arising from shunt complications (Lorber 1969) obscured the original purpose of the study. The fact that there was a considerable increase in the number of revisions required for the distal atrial catheter in these early shunts

and that a number of children with moderate hydrocephalus in the first week of life seem to achieve a balance between CSF production and absorption, without any shunt system,led to the conclusion that establishment of ventriculo-cardiac drainage in the first week of life was only justifiable in the child with very severe hydrocephalus.

Ventriculo-peritoneal drainage in the first week of life for the control of leakage of cerebro-spinal fluid from the back wound should be further considered; it carries less risk of immediate septicaemia in the presence of ascending infection from the back wound to the ventricles and the system is much less likely to be affected by the growth of the child than a system draining to the right atrium.

Hydrocephalus

Although the whole problem of the continuing care of myelomeningocele has been produced by the introduction of an effective method of control of progressive hydrocephalus, the multiple operations to maintain these shunts as shown in this series, the children who die of sudden occlusion of established shunts and the continual worry of the parents make these shunt systems one of the least desirable facets of the picture of the child with myelomeningocele. Other shunt systems, than the Holter one which was used in this series, do not seem to have any less complications (Cast et al 1969).

The basic complications are blockage due to relative movement of the upper and lower catheters and infection. It would seem that even with the figures in this small series the later a system is inserted the less likely is it to suffer from occlusion at the upper and lower end: delay in establishment of a shunt, however, must result in some brain damage and the risks have to be balanced. The use of the ventriculo-peritoneal shunt as a primary procedure theoretically might tide the child over a period of growth, so that a ventriculo-cardiac shunt could be established at a time when less differential shortening would occur between the catheter length and the distance between the valve and the right atrium, with the child's development. However, the complications of ventriculo-peritoneal shunt are devastating when they do occur, as in this

series where at least three of the peritoneal catheters perforated the alimentary tract and one at least produced not only an ascending infection but also an intussusception. We have also experienced in other cases intestinal obstructions due to adhesions around the distal catheter. Fixing the distal end of the peritoneal catheter above the liver or in the paracolic gutter has not proved a very successful method of avoiding complications. We still prefer the ventriculocardial shunt but possibly further trial with peritoneal or even pleural shunts should be made.

The figures in this series of nearly 14% infection of valve systems indicate the danger of this complication especially as 70% of the infected ones died. Apart from the systems colonised with staphylococus albus and those ventriculo-peritoneal shunts infected from intestinal perforations, at least nine children had infected systems which resulted from operative procedures. The length of time when tissues are exposed during an operation appears to play a part (Tsingoglou et al 1971) and this has been another reason for our reluctance to do elective revisions.

Tsingoglou and Forest (1968) have also been amongst those who have recommended regular elective lengthening of the distal catheter. In our experience elective revisions have not been without their troubles. There were only two in the series reported here but one of them suffered a left hemiparesis after the revision procedure and although this was only temporary such a serious complication following an elective procedure which possibly would be unnecessary, makes a surgeon very dubious about undertaking regular lengthening procedures. Yet to date there has been no controlled trial of elective procedures and such a trial is needed.

It is certainly clear that malposition of the tip of the lower catheter can lead to trouble not only when it is too high and allows thrombosis but also when it is too low and may encourage pulmonary embolus (Becker and Nulsen 1968). Sudden deaths from major pulmonary emboli have been reported (Noble et al 1970). The permanent presence of a foreign body in the blood stream cannot be regarded as anything but potentially harmful yet the removal of a shunt whose useful life has seemed to be long passed has resulted in sudden

death from acute intracranial hypertension presumably because the shunt has had minimal function. Again the surgeon must be very wary of unessential interference.

Perhaps the most important line of investigation is the search for a drug which will control hydrocephalus without implanted mechanical appliances. Glycerol, acetozolamide and isosorbide have been used (Cantone et al 1964, Wise et al 1968) but so far without very much evidence that they do any more than act by dehydration and in few cases has the reduction in CSF pressure been a prolonged one. Most of the work with these drugs has been on their effect on raised intracranial pressure in self-limiting conditions such as head injury. Foltz and Shurtleff (1968) showed that both isosorbide and acetazolamide produced an intital fall in pressure in hydrocephalic children followed by a rebound; Mealey and Barton (1969) suggest that though pressure may be reduced there is little effect on the progress of the hydrocephalus. In four children over one year old treated in Sheffield Children's Hospital acetazolamide slowed down the progression of hydrocephalus but all the children needed a shunt when the drug was stopped. A great deal more work is required on the use of drugs and this work is already under way in several centres.

The Urinary Tract

If one follows the principles of treatment already outlined, then each aspect of the myelomeningocele should be attacked in order to prevent its progression. Thus the progressive paralytic effects from exposure of the spinal cord are arrested by covering the cord at the primary operation, the progressive hydrocephalus is arrested by establishment of ventriculo-cardiac drainage and increasing foot and leg deformities corrected by appropriate orthopaedic treatment. Progression of the disease in the renal tract is far less obvious on clinical examination, however, and most practitioners concerned in the care of children with myelomeningocele have experienced the completely unexpected deterioration of the upper urinary tract in a child who appeared to be well under control but had a normal pyelogram one year and gross hydronephrosis the next. Not only may this apparent quite sudden dilatation occur but also the insidious destruction of renal tissue as a result of chronic pyelonephritis or even of repeated acute infections may lead to a degree of renal insufficiency of such a degree that even minor strains on the metabolism cannot be withstood: for example an apparently healthy child may be precipitated into uraemia simply by

the denying of fluids for four hours before a minor orthopaedic operation.

Because of the risks of deterioration and the probability in most cases of abnormal urinary control, early diversion of the urinary stream away from the potentially obstructed bladder to a bowel conduit has certain attractions in that it might avoid deterioration altogether. But urinary diversion using bowel conduits is a major procedure with many complications even in experienced hands (Cook, R.C.M., et al 1968) and it seems likely that later complications are going to multiply particularly with regard to hypertension. In a group of ileal loop diversions reaching late teens and early twenties, two girls died of eclampsia during pregnancy and almost all had a degree of hypertension (Rickham 1971). Even if the procedure were trouble free, the 75 children who attained acceptable dryness at the age of five in this series discourages embarking on a major procedure in all cases - even if "acceptable" dryness at 5 is no longer acceptable at 15.

The capacity of recovery of the kidneys is enormous in the growing child. A few children have been shown to have hydronephrosis in the neonatal period and yet developed normal upper renal tracts at a year of age; this spontaneous improvement has not been explained but is probably due to temporary overcoming of relative outflow obstruction and should not lead the clinician into any complacency about early hydronephrosis. More reliably, and perhaps more importantly, provided the twin pronged attack on the kidney by obstruction and infection has been removed, the nephrons in a growing child are capable of hyperplasia which is not seen in adults; thus even in a child who has had quite extensive destruction of nephrons, considerable recovery may be hoped for after a "decompression" procedure, provided infection can be kept under control. (Emery 1971)

It thus seems reasonable that a calculated risk should be taken, and that surgical procedures on the urinary tract should be undertaken only when there are positive indications, in the same way as antibiotic therapy is applied vigorously when indicated but not as a prophylactic procedure. But in the same way as a close watch is kept for developing infection, some method of very early recognition of departure from the normal is required.

It is believed by some authors (Smith, 1965, de Jouge et al 1969) that all the

progressive upper urinary tract disease in myelomeningocele is not due to bladder outlet obstruction; they suggest that there are intrinsic abnormalities, perhaps of neuro-muscular origin in the renal pelvis, the ureter or at the ureterovesical junction. We have seen the higher incidence of associated urinary tract abnormalities in this series than in otherwise normal children and such anatomically malformed kidneys as horseshoe kidneys or kidneys with pelviureteric obstruction are more liable infection; but no intrinsic abnormality in nerve fibres or ganglia has been shown in the ureters of children with spina bifida (Forbes et al 1969) so there is little pathological justification for blaming upper urinary tract dilatation on anything other than the known imbalance between detrusor activity and bladder outlet resistance: even when vesico-ureteric reflux has not been shown a high intravesical pressure could account for upper urinary tract dilatation.

The success of a conservative approach to treatment and avoidance of progressive damage to the upper urinary tract will depend, therefore, on the very early recognition of imbalance between detrusor and sphincter mechanisms. The routine management in these children has already been described. Further information has been gained by experimental study of intravesical pr ssures. My first attempt was an unpublished preliminary investigation involving the study of bladder pressures carried out in 1967; thirteen children were studied of whom three were excluded, two for technical difficulties and one because he was an older child. Ten babies with open myelomeningocele were studied at the earliest possible moment in their lives at the time of the back closure when they were under 24 hours of age. In all of them (except two with very extensive limb paralysis) some sign of bladder tone was demonstrated: this took the form of a sustained rise in bladder pressure to between 5 cm. to 15 cms. H_2O, maintained for two to three minutes. These were rather crude measurements using a transducer measuring water pressure through a catheter passed into the bladder per urethram. Their main value was in demonstrating that whilst bladder pressure changes could certainly be recorded, they were never produced by stimulation of the neural plaque of the myelomeningocele in the same way as plaque stimulation would often produce contraction in lower limb muscles which otherwise appeared paralysed. Assessment of the type of bladder dysfunction was therefore no more successful by this investigation than by attempted correlation to the spinal lesion. (Fig. 6).

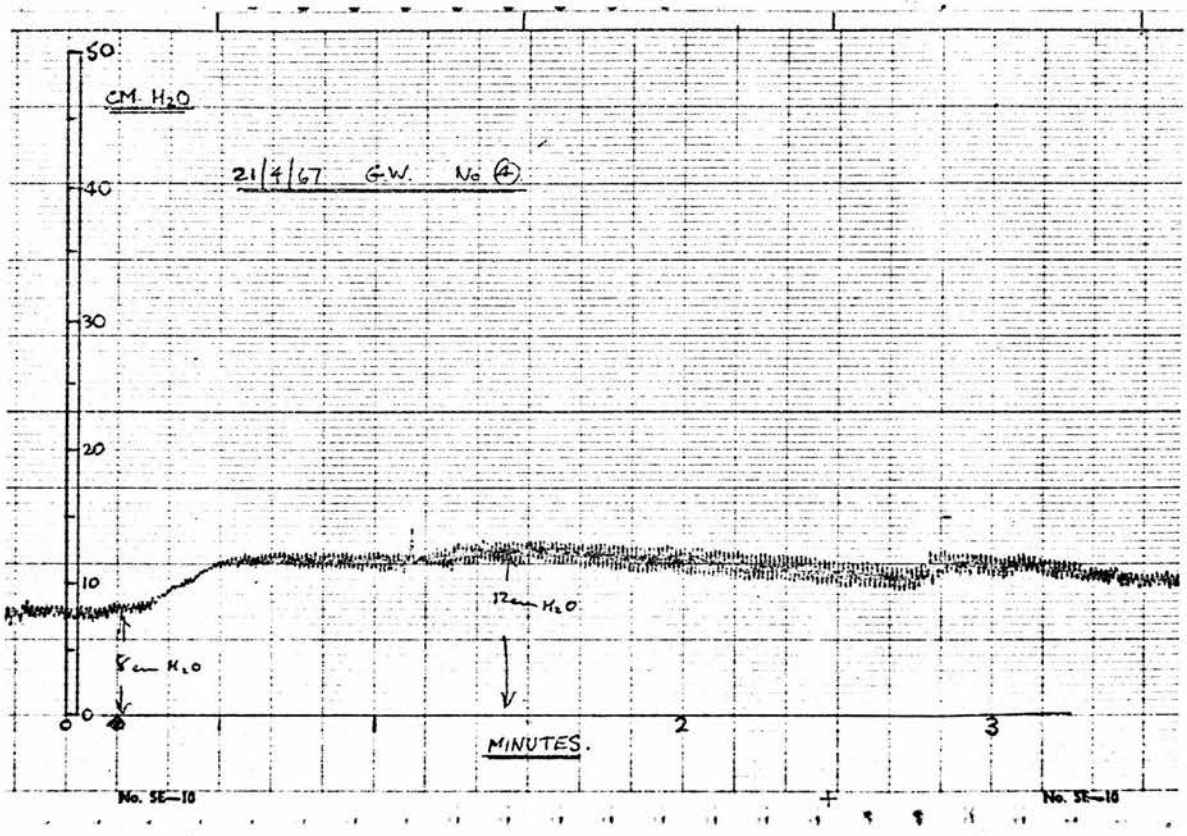

Figure 6 : Bladder pressure curve at time of back closure

Later development of more sophisticated methods of bladder pressure studies (Pekarovic et al 1969) in our research laboratory have given a good deal more indication of the general patterns of malfunction: the equipment used has involved the continuous filling of the bladder through a suprapubic cannula recording bladder pressure through the same line. Bladder filling is constant at 2 mls. per minute and is measured by recording of the filler bottle weight by a strain gauge; urine outflow per urethram is collected in a burette and recorded by a second strain gauge and each drop of urine from the external urethral orifice is registered by a bead thermistor. Information is thus gained about the nature of detrusor contractions and the effectiveness of bladder emptying. The types of curves could be placed in two groups.

(a) Curves with detrusor contractions:

In this group the most common pressure curve is that which shows easily recognisable detrusor contractions at intervals of $\frac{1}{2}$ - 2 minutes at an inflow rate of 2 mls. per minute. The contractions produce a pressure of 60 - 70 cms. of water and are accompanied by small streams of urine of 2 - 8 mls. In this group the outflow of urine is usually free and hence these contractions are sufficient to empty the bladder completely, or to leave only a small amount of residual urine, not more than 10 mls. (Fig. 7).

Sometimes the detrusor contractions are not strong enough reaching only 40 cm. of water and do not empty the bladder completely, even if there is no obstruction in the bladder outlet. Here the residual urine may be as much as 40 mls. (Fig. 8).

When some obstruction to the bladder outlet is present the detrusor contractions increase to 100 - 150 mls. of water and even higher. Usually in these cases there is a baseline pressure of 30 - 40 cm. of water, which is higher than the 10 - 20 cm. in those where there is no obstruction to the bladder outlet. The duration of contractions is longer and the frequency decreased. In spite of the high pressures reached the bladder is not effectively emptied and a large residual of urine of up to 100 mls. is always present. (Fig. 9).

All these types of activity are pathological; normal children were not studied, though by comparison with children with minimal abnormalities we believe

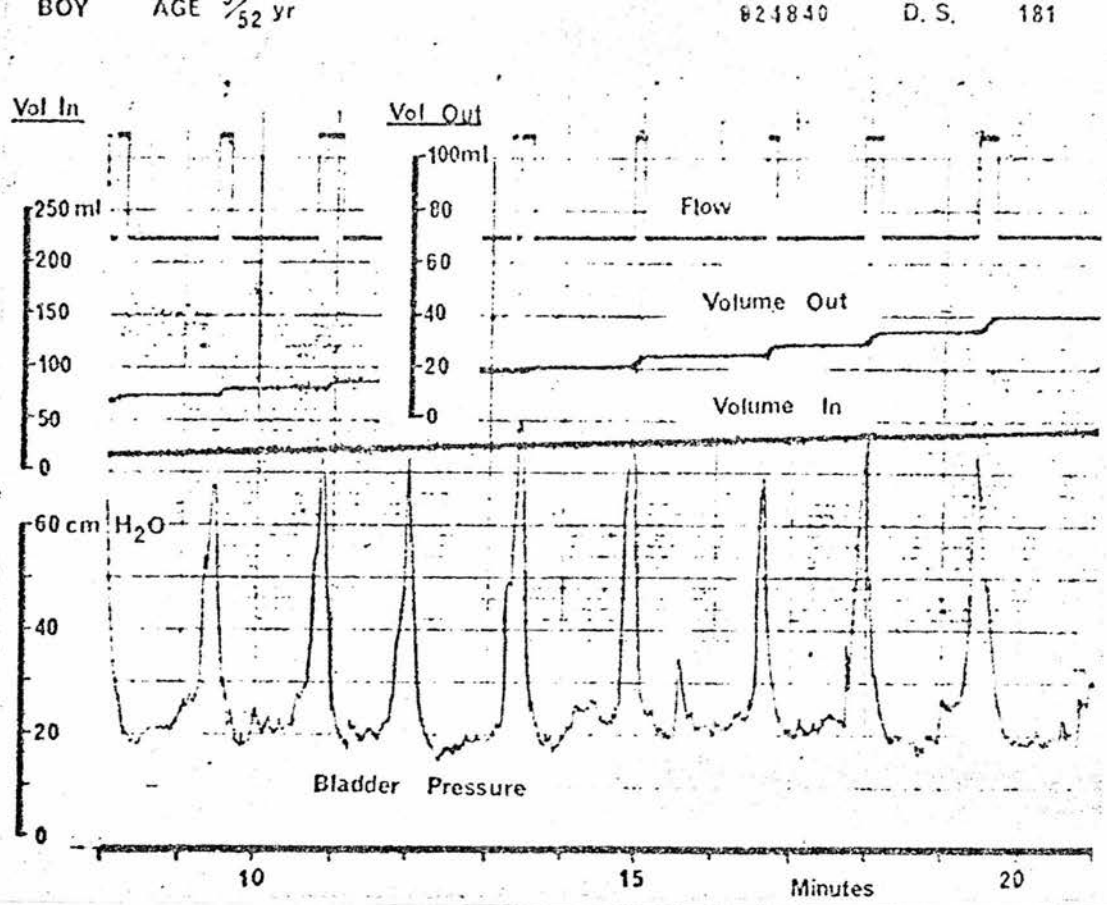

Figure 7: Normal pressures but excessive frequency: contractions empty bladder.

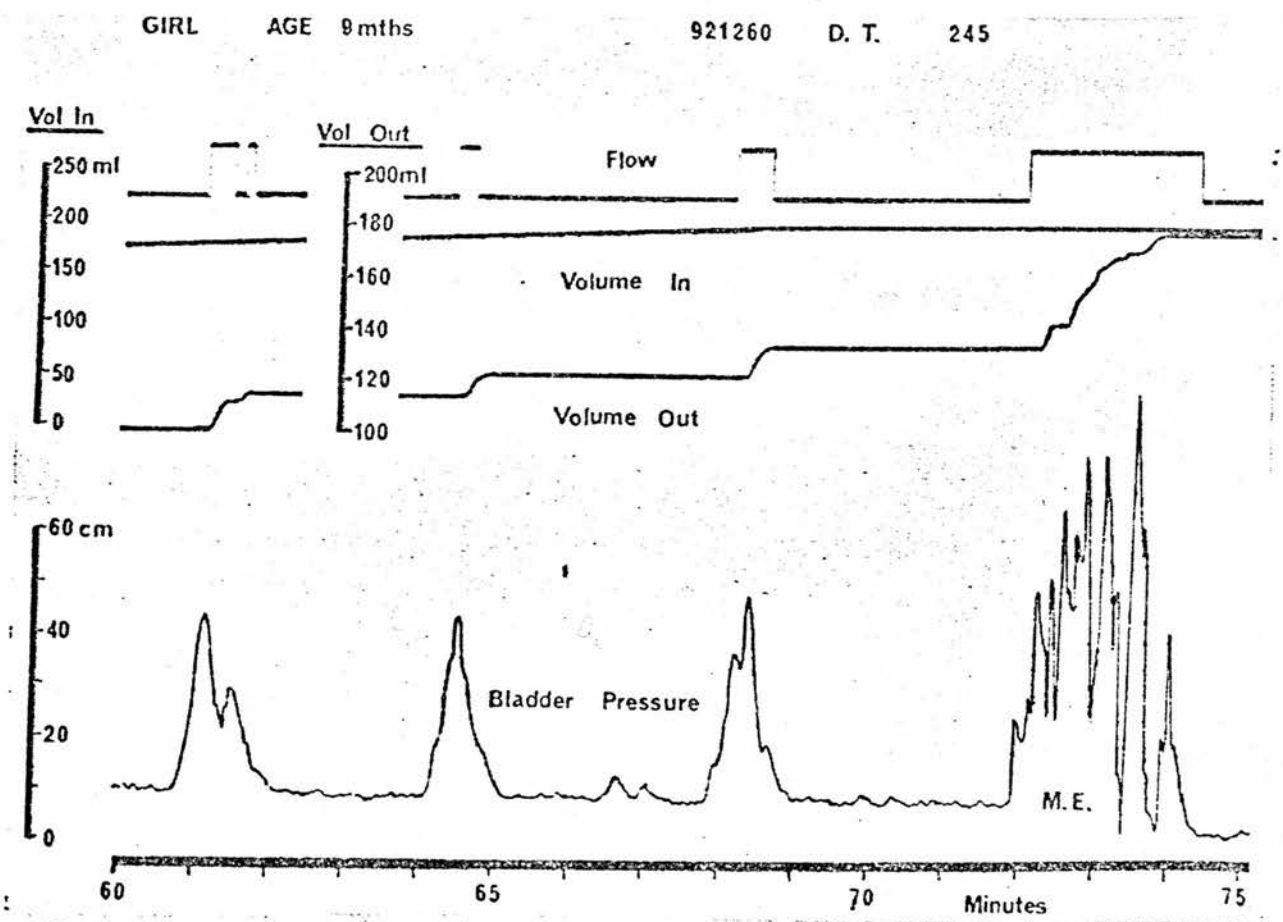

Figure 8: Low pressure contractions do not empty bladder but suprapubic pressure (M.E.) completes emptying.

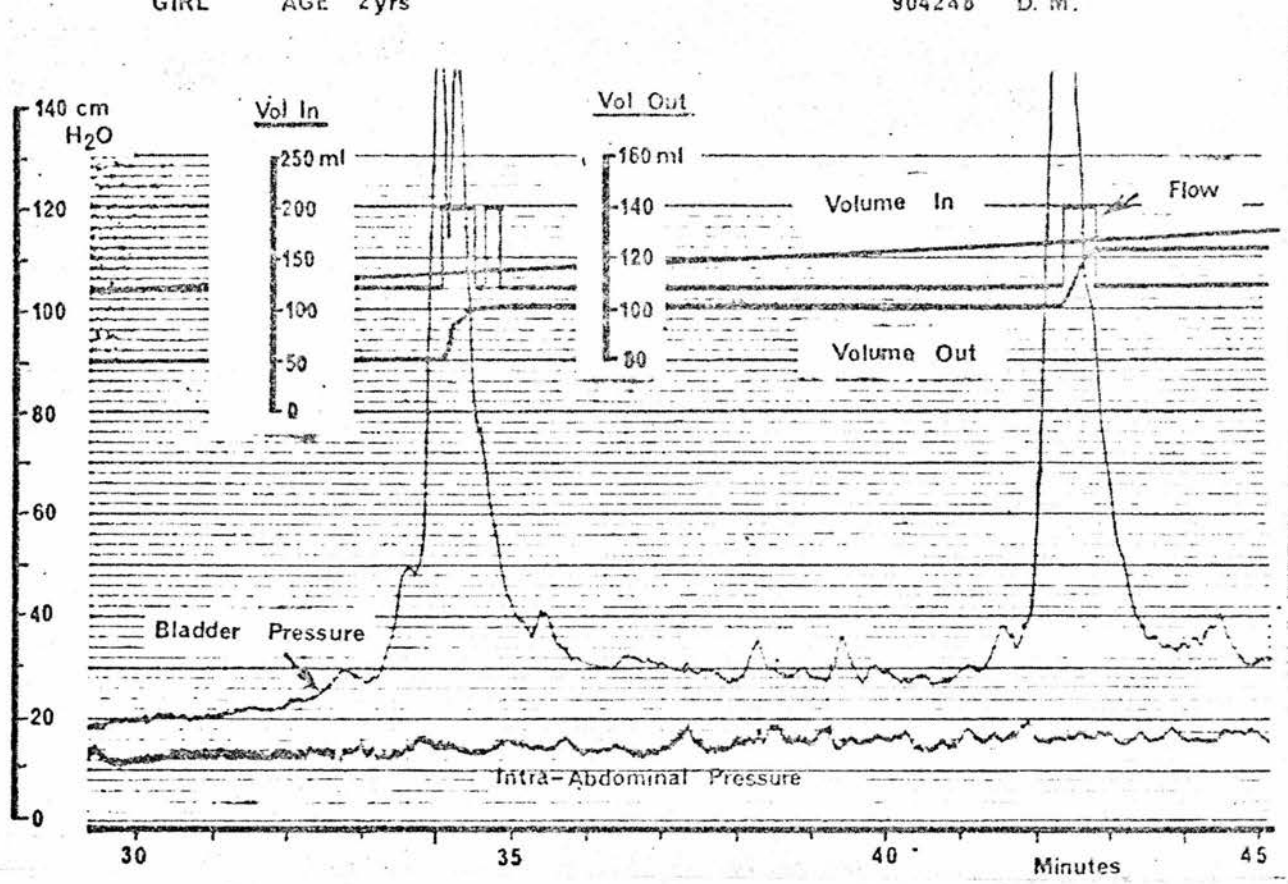

Figure 9: High pressure contractions reaching 150 cms. H_2O but failing to empty bladder: outlet obstruction.

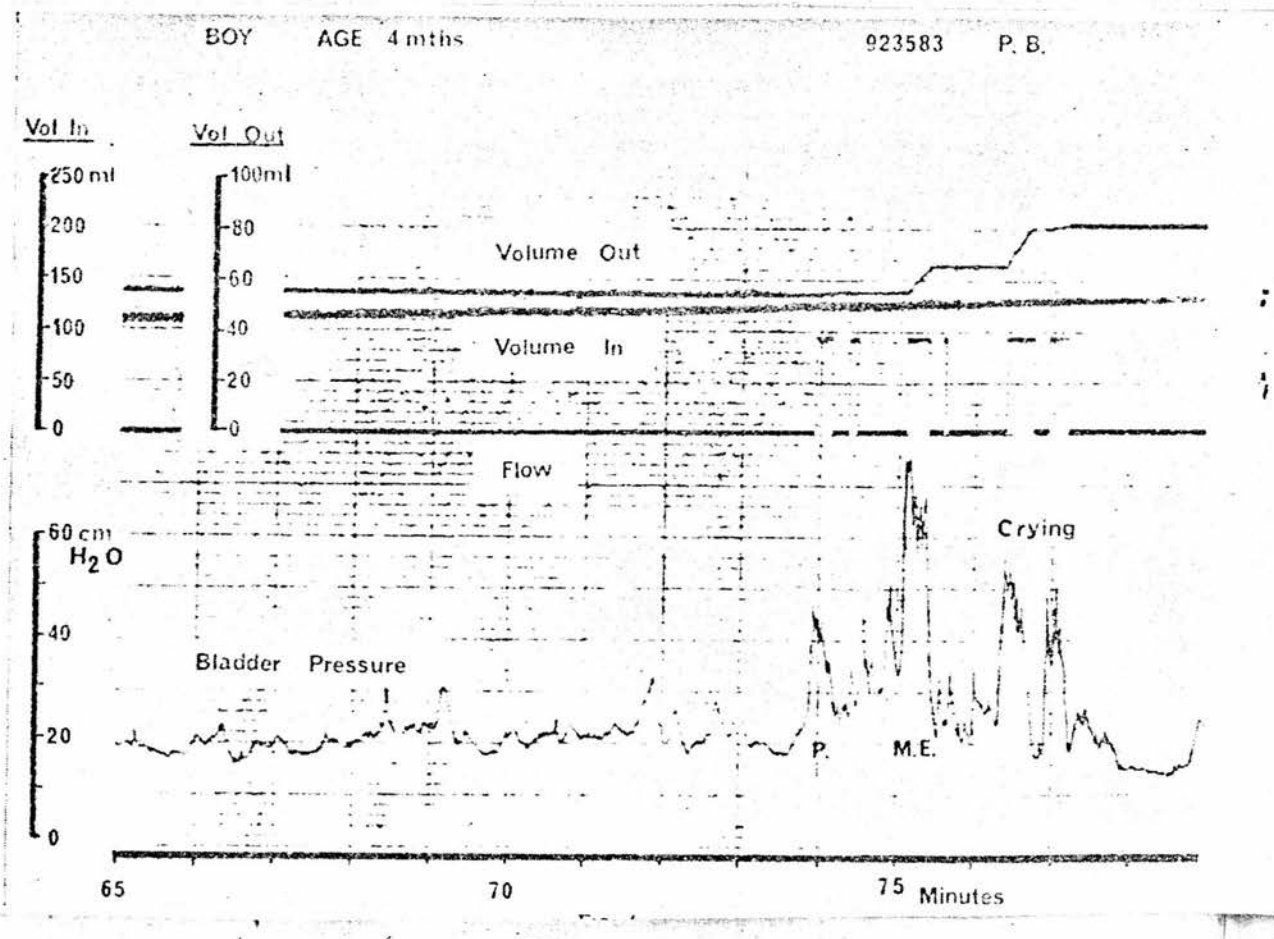

Figure 10: Atonic bladder without contractions: palpation (P) produces slight stream and suprapubic expression (M.E) empties bladder.

that the normal bladder in the child will produce a pressure of 60 - 70 cm. of water and each contraction will produce a stream of 30 - 40 mls. of urine. The bladder is emptied completely and there is no dribbling.

(b) Flat curves without detrusor activity:

In this group the bladder pressure curve is almost flat at a level of 10 - 20 cms. of water showing only slight changes of 4 - 8 cms. caused by extravesical pressure. No detrusor contractions are observed and emptying of the bladder of this nature occurs only during straining by moving, crying, laughing or coughing. Complete emptying of such a bladder is unusual and most cases had a residual urine often of a very large amount, over 200 ccs.

Where there is obstruction of the bladder outlet in patients with flat curves, the baseline pressure is perhaps 30 - 50 cm. of water and there is a larger amount of residual urine.

These intravesical pressure studies have clearly had their value in defining the nature of bladder dysfunction: but the main way in which they can play a part in the clinical management of the child with myelomeningocele is by indicating the child in whom urinary tract deterioration is likely before any of the other investigations would indicate this. For example, if bladder pressure studies were to show high intravesical pressure even though there were no residual urine then one could conclude that an element of outflow obstruction was being overcome by detrusor hypertrophy, and one might also anticipate that the bladder was likely to fail in time and that later investigations would demonstrate the presence of residual urine. This very situation has occured on a few occasions and thus bladder pressure studies could be claimed to be the most sensitive of all metnods of recognising the first evidence of an upper urinary tract that is likely to fail.

The equipment, however, is expensive and investigations time consuming, and the simple estimation of the presence of redidual urine after micturition or after manual expression gives very good evidence of ineffective bladder emptying; intravenous pyelography and micturating cystourethrography will complete the investigations required for making a decision as to the nature of surgical intervention required.

Bladder pressure studies have also been helpful in indicating those cases which can be helped by suprapubic bladder expression and those which can not. If there is no outflow obstruction and normal detrusor contraction the bladder will remain empty and expression is unnecessary. If there is no outflow obstruction and low pressure or absent detrusor contractions then manual expression will help. In the pressure of outflow obstruction manual expression may be unsuccessful; in all bladder studies the pressure obtained by manual expression was recorded and usually reached 80 - 100 mls. water. This is far short of 150 mls. produced by an active detrusor muscle in the presence of outflow obstruction (Figs. 9 and 10) and will not empty the bladder. Indeed manual expression may do harm in the presence of outflow obstruction by encouraging vesico-ureteric reflux; loin pain can be produced in some children by suprapubic pressure: admittedly in the child with an active detrusor and outflow obstruction more damage will be done by the detrusor contractions than the manual expression but in an atonic obstructed bladder supra-pubic pressure should be avoided.

Similarly isotope renograms play a useful though not essential part in investigations. Their main value is in the comparison of serial examinations; (Johnston et al 1967) which can be frequently repeated because of the low radiation dose compared with radiography; evidence of change would lead to further radiological studies.

The type of surgical intervention again will depend on the stage to which the disease has progressed. One would hope that the discovery of inadequate bladder emptying has been made before there is evidence of upper urinary tract dilatation, in which case surgery will be aimed at widening the bladder outlet. In the series seven children had Y.V. plasty procedures on the bladder neck; this could only be regarded as a temporising procedure, as shown by the poor results in six of them. In fact later investigations including more accurate cystourethrograms have shown that it is only rarely that the bladder neck is the site of the obstruction and much more commonly the narrowing is at the level of the external sphincter which cannot effectively be reached in the transvesical procedure of Y.V. plasty. Attention has therefore been directed to the external sphincter.

Pudendal neurectomy in the one case in this series was disappointing; it will only be effective when there is actual spasm in the sphincter. Stark (1968) showed that when there was a positive anal reflex electromyography revealed active obstruction at sphincter level; here pudendal neurectomy might be expected to help. In most cases, however, the obstruction is not active and there is no positive anal reflex and a deliberate division of the sphincter is necessary. Transurethral resection sphincterotomy seems ill designed because the instrument which cuts the sphincter must first cut the transitional urethral epithelium and there may well be extravasation of urine. A direct attack on the sphincter alone seems a much more attractive approach and initial results achieved by dilatation of the urethra in girls and a perineal approach sphincterotomy in boys (whose anterior urethra is too narrow to allow adequate posterior urethral dilatation) have given encouragement to this method of management (Zachary 1971).

Cutaneous ureterostomy, though it has achieved some remarkably successful results in the series, is an operation that should never be done if patients are being adequately followed up. It is only the dilated ureter that can be fashioned into a suitable skin spout for drainage, thus if a ureterostomy is possible then the upper urinary tract has been allowed to dilate and urinary diversion is too late.

Urinary diversion, using a bowel conduit, is therefore the treatment of choice when bladder outlet procedures have failed to allow the continued free flow of urine. There seems little doubt, however, that more and more of the children with bowel conduits are going to have further problems: a considerable number of them require revisions of their loops; three in the series had already been revised by the time they were five. Continued deterioration of the upper urinary tract after a loop diversion may be explained by the fact that the upper urinary tract was badly dilated at the time the loop was made or by technical faults in the procedure such as a long kinked loop or stenosis at the junction between ureter and bowel or between bowel and skin. A number, however, have been found to deteriorate without these obvious explanations and it is most disturbing to find deterioration in a normal upper urinary tract in a child whose diversion was made on purely social grounds. In these it may be that high pressures within the bowel conduit have produced a functional

obstruction (Pekarovic et al 1968) - and it has been notable that the pressures within sigmoid loops tend to be higher than in ileal loops. Since the purpose of the conduit is to provide free, unobstructed drainage of urine these studies might suggest that a loop of ileum is preferable to a loop of colon. Mogg (1965) strongly upholds the sigmoid colon as a satisfactory bowel conduit and it may be that his technique of making an anti-reflux anastomosis between ureter and sigmoid is the explanation of his success; certainly experience would indicate that some effort should be made to protect the kidneys from the effects of a high loop pressure when colon is being used for the conduit.

Two other points should be mentioned in the consideration of the urinary tract. First with regard to infection; it is now well recognised that infection in the urinary tract is a sign of serious underlying pathology and not something to be treated merely with appropriate antibiotics as and when it occurs. Certainly infection should be vigorously treated after identification of the organism and its sensitivities; but at the same time infection of the urine should be an indication for a full investigation, or re-investigation of the urinary tract. Prophylactic antibiotics have not been shown to have much value, though there might be a place for them in those cases where no obstruction to the urinary outflow can be shown yet the kidney remains the site of chronic pyelonephritis; an example would be in the case where an anatomically satisfactory urinary diversion has been made in a child with irreversible upper urinary tract dilatation.

Second, the use of electrical stimulators. There may be a place for electrodes implanted in the pelvic floor to control wetness in those rare cases where frequent low pressure contractions keep the bladder empty (Caldwell 1967) but this situation is very rare. Electrical stimulation to increase detrusor activity depends on many things including wide stimulation of an effective muscle and this is really unlikely to be present. There does not seem to be much scope for implanted electrodes and stimulation from an external source except in a small number of patients with minimal lesions. More research work is required in this field.

When one looks again at the results achieved in the 200 cases in the series one must consider carefully the 57 children who at the age of five are neither dead nor near normal. It has been suggested by the paediatrician who works in our team (Lorber 1971) and by others (Stark 1971) that the lives of children are being preserved who will be a misery to themselves and to their families; who will suffer continuing pain and disability until they eventually die of the complications of the disease. It cannot be denied that all of the survivors will be paralysed to some degree and many of them severely; they were all paralysed to some degree when they were born and many of them severely. Nor can it be denied that the burden of care of the severely handicapped child is one that could be gladly shed: treatment in the knowledge from the outset that the result will be a handicapped person is a burden for doctor, patient and family.

Selection of patients for treatment essentially means selection of some patients for non-treatment, in the hope that they will die.

When one considers that the whole principles of treatment are directed to the preservation of function then non-treatment must mean the non-preservation of, or loss of, function. In many cases, however, it does not mean loss of life. A fundamental fault in the arguments in favour of selection of patients for treatment has been that which was put forward by the opponents of Newbigging in 1834 - that the alternative to treatment was early death. To this day the same alternative is often given to parents,"would you like your baby treated and exposed to all the complications of this congentical deformity or would you prefer to allow nature to take its course?". A child with an unclosed back may develop meningitis and die within a few weeks of birth; a child with uncontrolled hydrocephalus may die within a few months and a child with unrelieved urinary outflow obstruction, back pressure and chronic infection may die in the first few years of his life. None of these could be called a "natural" death and all are likely to cause prolonged suffering to the child and his family. Basically the condition is not immediately lethal like, say, congenital oesophageal atresia or intestinal obstruction where death in the untreated case is certain and early. Thus the only

logical alternative to undertaking the treatment of the child is not to withhold treatment but to kill the child deliverately, an action unlikely to be taken by a doctor who has been trained to cure.

Death in the untreated case is therefore not certain. Nor indded is it certain that the eventual result in the severely affected child will be as bad as appears likely at birth. Expert examination may indicate the minimal handicap which may be expected (Stark 1971) but such axamination demands the admission of the child to a specialised unit where it may be difficult to refuse specialised care. The classification in this series was designed to be simple enough for any person to group the child, assuming that while the apparent level of the lesion might not be its neurological level, at least the extent of the lesion would bear some relationship to the final result. This indeed was shown to be true in the series, the more extensive lesions carrying a higher mortality rate and poorer overall result. But the bizarre histology of the local lesion on the back presumably accounts for the surprises experienced by clinicians and even specialist paediatric neurologists when a child with a severe lesion at the age of five is a great deal better than had been anticipated. Five of the eleven survivors with the most severe lesions fell into the least handicapped category at the age of five.

To withhold treatment from the newborn child one must be practically certain of two things, first that he will die if untreated and second that treatment will only result in the survival of a very severely handicapped child. We can be certain of neither of these at the time the child is born and we must therefore undertake treatment in all cases. There are perhaps two situation where immediate active treatment can justifiably be delayed: first, when the child has evidence of severe intracranial damage and seems likely to die within twenty-four hours; and second, when there is an extensive lesion and complete limb paralysis where the difficult closure of the back might well increase the chances of infection because of would breakdown. The latter is substitution of conservative for operative management, not non-treatment.

The possibility of infection is present in all cases whether treatment has been operative or conservative and local infection of the back wound means a

potential meningitis and ventriculitis. Such infection carries with it a very high mortality as shown by the 29 who died in this series in spite of vigorous treatment. A proportion of the survivors will have suffered deterioration of brain function as a result of the infection and in those with hydrocephalus the necessary delay in its treatment whilst the infection is being treated with antibiotics may result in further deterioration. The surgeon may be in the position of seeing a child suffering from rapidly increasing hydrocephalus and being unable to establish a shunt system because of the certain knowledge that the system would become infected as soon as it was inserted. A similar situation was seen in valve systems that had to be removed because of infection; there was an inevitable delay whilst treating the infection before a new system could be inserted. The high mortality of untreated ventriculitis and the poor prognosis for the treated case, especially where hydrocephalus is also present, make this a situation where the clinician might give some thought as to how much is to be gained by treating the child. Once more one has to face the fact that not every untreated child will die and not every treated child will be severely affected, and the clinician can hardly be unaffected in his decisions by the presence of the severely handicapped survivor who was not treated.

Possibly the only situation where death is certain if treatment is delayed is in the child with acutely raised intracranial pressure due to a blocked ventriculo-atrial shunt system. Here the child is frequently admitted to hospital and one child in the series died in this situation. If such a child were mentally defective and severely paralysed it might seem unjustifiable interferance for a surgeon to attempt to relieve the situation by revising the shunt system. In practice, however, the parents of such a child are almost invariably anxious that every possible measure should be taken to save his life. The general public rightly assumes that the medical profession will do all it can to treat the illness of a patient; if a decision is made to avoid therapy then a signed declaration of agreement to this decision is probably as important as the signed consent form for operation.

No matter how much the clinician might desire to avoid taking the decision to treat or not to treat, the only way in which that decision could be taken for him would be by direct instruction on purely financial grounds. If limitations are to be placed on the extent of medical treatment on the grounds of limitation of available funds, then that is a decision for the general public to make. The doctor

who is employed by them is bound to follow their instructions. However, the situation of the public demanding a ban on the treatment of handicapped children is unimaginable and the decision is therefore bound to remain with the doctor.

No agreement has been reached amongst the medical profession on the question of selection of cases for treatment. Controversy is understandable and probably healthy, but it is unfortunate that two recent papers recommending selection (Lorber 1971 and Stark 1971) reached the national press from scientific journals (photocopies of some of the comments are in Appendix II). It was suggested in one editorial that parents might be concerned about bringing their child to a hospital which had been reported as advising that severely disabled children should be allowed to die. In fact in Sheffield since this publicity some patients have asked for the name of the consultant treating their child for fear that every effort would not be made to save his life, and this was a child with a totally different congenital anomaly from myelomeningocele. I believe that such undermining of public faith in the medical profession is a very serious consequence of a declared belief in selection and that this must affect the decision of the clinician to adopt these measures, or at least to publish them.

Whilst under no circumstances should non-curative treatment be undertaken if it is likely to cause pain or discomfort, I believe that to leave the child untreated is to evade one's responsibilities. If the results of treatment of a condition are bad then we must try to improve them, not give up altogether. Our aim in the treatment of all congenital abnormalities which cannot be completely cured should be the same as our aim in the treatment of any other patient, young or old, with incurable disease; major disabilities should be controlled as far as possible, general health should be supported. Palliative treatment is indicated for the relief of pain and discomfort. In the child this may mean treatment that does little more than make nursing care easier, but in many cases the end results are surprisingly good in the widespread abnormalities associated with spina bifida.

Wilt thou also destroy the righteous with the wicked?

Genesis 18, 23.

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APPENDIX I

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
PALLISTER, Mandy 875480	29. 6. 62.												T/L/S	1			10/52	
URSE, Tracey 874915	31. 5. 62.											S	L/S	34		✓		
LATER, Elaine 874964	1. 6. 62.								✓	2		S	L/S	26	✓			
WILSON, Mark 875019	5. 6. 62.												L/S	15			✓	Ventriculitis - died aft 14 weeks treatment. N.B. Later closure
PLUMPTON, David 875023	4. 6. 62.				✓						✓✓✓✓✓	O	L/S	54	✓	✓		
FREEMAN, Paul 875132	8. 6. 62.	✓					✓		✓	✓	✓	S	L/S	40				One nephrectomy One hydronephrosis at 5
BROOKS, Martin 875146	9. 6. 62.					✓					✓✓✓✓	S	L/S	11	✓	✓		Craniostenosis
BRADFORD, Karen 875178	12. 6. 62.								✓		✓✓✓✓ ✓✓✓	O	L/S	23				Hydronephrosis Controlled
TERRY, Geraldine 875338	20. 6. 62.								✓		✓✓✓ ✓✓✓	S	L/S	16				A lot of stoma trouble Controlled hydronephrosi
HATTIE, Amanda 875441	23. 6. 62.						✓	✓✓	✓		✓✓✓✓	O	T/L	78	✓			Nephrolithotomy Pyelolithotomy Pyelolithotomy Pyelolithotomy Lithotomy

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
BELL, Rosalyn 875575	30. 6. 62.										✓	S	T/L	15	✓	✓		
WATSON, Trevor 875682	3. 7. 62.										✓✓	O	S	6	✓	✓		Large head at 5 yrs.-21½
McMULLEN, Susan 876026	19. 7. 62.												L/S	2			✓ 2/52	Peritonitis Ruptured bladder
HAGUE, Heather 876061	20.7. 62.	✓	✓										T/L	13			✓ 14/12	Renal failure
WHITTAKER, Julie 876084		✓	✓✓		✓						✓✓✓✓ ✓✓	S	T/L/ S	47	✓	✓		
WILROY, Sharon 876162	25. 7. 62.										✓✓	S	L	32	✓	✓		
LAY, Darren 876197	27. 7. 62.												T/L	14			✓ 13/12	Coroner's Report: Gastro enteritis. Solitary kidn Hydronephrosis.
MOSS, Tracey 876228	27. 7. 62.				✓						✓✓✓✓	O	T/L	30	✓	✓		
McDONALD, Linda 876234	28. 7. 62.	✓	✓✓								✓✓✓✓ ✓	S	L/S	33		✓		Awaiting diversion
COLEMAN, Diana 876510	14. 8. 62.										✓		L/S	6			✓ 6/12	Probably respiratory

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
IMMS, Steven 876668	21. 8.62.											S	T/L	13				Unilateral paresis Hydronephrosis at 5
ARTSHORN, Ann 876798	27. 8.62.	✓	✓✓ ✓										T/L	13			✓ 20/12	Urinary infections had occurre Said to have multiple pulmonary embolisms
ARRISON, Andrew 876907	1. 9.62.												T/L	2			✓	Ventriculitis at 2/52
INCHBECK, Timothy 876912	1. 9.62.	✓					✓						T/L	7			✓ 15/12	Early X-ray (R) hydronephrosis Pyonephrosis.
OLOWSKI, Alison 877069	10. 9.62.										✓✓✓ ✓✓	S	T/L	17	✓	✓		
AYNE, Tina 877295	20. 9.62.												L/S	3			✓	Meningitis 3 Ventriculitis weeks
IGGOTT, Vivian 877355	23. 9.62.												T/L	2			✓ 2/52	Ventriculitis No post mortem
EARCE, David 877358	23. 9.62.				✓							S	T	3	✓	✓		
ILLEY, Dawn 877417	25. 9.62.												T/L	3			✓ 3/52	Ventriculitis Hydronephrosis

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
FOSTER, Mark 877522	28. 9.62.										✓	0	S	8	✓	✓		
SWIFT, Beverley 877647	4.10.62.	✓	✓✓✓	✓								0	T/L	22	✓		✓ 5 yrs.	Diastematomyelia Re-exploration Renal failure
EDWARDS, Michael 877756	10.10.62.	✓	✓✓✓										L	14			✓ 13/12	Renal failure
ADAMS, Raymond 877779	11.10.62.	✓	✓✓✓✓ ✓✓✓		✓						✓	S	L/S	35	✓	✓		
DODDS, Alan 877918	17.10.62.	✓		✓								0	T	8	✓			Definite myelomeningocele but minute plaque without response to stimulation
BOWYER, Annette 877936	18.10.62.								✓			0	T/L/S	37	✓			Renal stones Hydronephrosis
WRIGHT, Patrick 878397	9.11.62.	✓	✓✓										L	4				Believed well. Follow up incomplete. Family have mov to Ruislip
LEADER, Rosamund 878449	12.11.62.	✓	✓		✓							0	T	25	✓	✓		Later social diversion
BENNETT, Elizabeth 878524	15.11.62.												T/L/S	1			✓ 1/52	Microcephaly Pierre Robin

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
OSTLER, Ian 878553	17.11.62												T/L	2			✓ 2/52	Intraventricular haemorrhage
JOHNSON, Dale 878694	23.11.62	✓	✓✓ ✓✓										T/L/S	16			✓ 1.11/12	
REYNOLDS, Jacqueline 878811	29.11.62								✓		✓✓	S	L	23	✓	✓		
BETTS, Andrew 878849	30.11.62	✓									✓✓ ✓✓	S	L/S	34				Diastematomyelia (dermoid) Re-exploration. Hydrocephalus controlled
MILLWARD, Jill 878584	6.12.62												L/S	2			✓ 2/52	Craniotomy by obstetrician P.M.multiple brain haemorrhage
WHITEHEAD, Jacqueline 879171	17.12.62										✓✓ ✓		T/L/S	2			✓ 2/52	P.M. Septicaemia Gross hydronephrosis
FISHER, Mandy 879221	24.12.62				✓							0	L/S	17	✓	✓		
DAVIES, Angela 879268	23.12.62												L/S	6			✓ 3/12	Died of gastro-enteritis elsewhere Mongol

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
CARTER, Denise 879323	29.12.62.	✓	✓✓						✓		✓✓✓	S	T/L	34	✓			Hydronephrosis controlled
SIMPSON, Christine 879543	13. 1.63.	✓	✓✓					✓					T/L/S	17			✓ 14/12	Renal failure
EANSWORTH, Kevin 879571	14. 1.63.	✓	✓✓✓ ✓			✓						O	L	16		✓		
LINDSAY, Gail 879673	19. 1.63.												T/L	10			✓ 10/52	Ventriculitis
LEONARD, Pamela 879679	19. 1.63.			✓							✓✓✓ ✓	O	L	16.		✓		Said to have considerable paraplegia at birth but recovered
FLEMING, Susan 879802	25. 1.63.												L	8			✓ 2/12	
OLDHAM, Margarita 879805	26. 1.63.	✓	✓✓ ✓										T/L	16			✓ 20/12	Ventriculitis
CASEY, Dean 879879	29. 1.63.	✓	✓✓										T/L	30			✓ 20/12	Ventriculitis

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
WALKER, Angela 880001	6. 2.63.												T/L	5			✓ 5/52	P.M. Ventriculitis Single hydronephrotic kidney
STOREY, Eamon 880020	7. 2.63.	✓	✓✓										L/S	17			✓ 4/12	P.M. Septicaemia (valve was removed for sepsis)
DOLBY, Raymond 880078	10. 2.63.	✓	✓✓ ✓✓			✓					✓	S	T/L	24		✓		Low I.Q.
EAGLEN, Gary 880123	12. 2.63.	✓				✓						S	T/L	22	✓	✓		
SLACK, Dawn 880505	3. 3.63.												T/L	1			✓ 2/365	Inhalation meconium (birth injury?)
WILSON, Dawn 880590	6. 3.63.												S	8			✓ 20/12	Fibrocystic disease
FRANCIS, Lesley 880605	7. 3.63.				✓						✓	S	T/L	30	✓	✓		
MARSH, Angela 880638	8. 3.63.	✓			✓						✓✓✓	O	L	6	✓	✓		
LEADLEY, Tracey 880753	13. 3.63.				✓							O	L	17	✓	✓		Had re-exploration of back

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
GORDON, Jean 880690	12. 3.63.												L/S	1			✓ 10/365	Bilateral hydronephros Respiratory infection Wound infection Had steroids
NIGHTINGALE, David 880754	13. 3.63.	✓		✓							✓✓✓ ✓✓	S	L (high)	4	✓	✓		
MARSDEN, Jayne 882126	13. 5.63.								✓		✓✓	O	L/S	9	✓	✓		
MOFFETT, Anna 882151	14. 5.63.			✓							✓	O	L	3	✓			
HARRISON, Lorraine 880799	15. 3.63.				✓						✓	O	S	5	✓	✓		
HOLMES, Bryan 881027	25. 3.63.	✓	✓				✓		✓	✓	✓✓✓ ✓	S	T/L	44	✓			
BATTY, Stewart 881188	2. 4.63.	✓	✓✓ ✓✓			✓					✓	S	L/S	24		✓		Very low I.Q.
WILLERTON, Sarah 881250	4. 4.63.												L/S	<1			✓ 5/365	Ventriculitis

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
BOOTHROYD, Alison 881298	6. 4.63.												L/S	5			✓ 5/52	Ventriculitis
WOODHEAD, Keith 881324	7. 4.63.	✓									✓✓✓ ✓	0	T/L	14	✓	✓		Duplication ileum
LONDON, Susan 881579	21. 4.63.												T/L	8			✓ 2/12	Bilateral pneumonia
HOROBIN, Joanne 881615	24. 4.63.	✓	✓		✓		✓					0	L	12	✓	✓		
DAFT, Mark 881739	27. 4.63.			✓								0	L/S	4	✓			
BOOTH, Karen 881736	27. 4.63.	✓	✓		✓							0	L/S	10	✓	✓		Diastematomyelia
THOMSON, Paul 881778	29. 4.63.	✓	✓✓					✓	✓		✓✓✓	S	L/S	24	✓			Hydronephrosis uncontrolled
KINGSTON, Peggy 881780	29. 4.63.								✓	✓	✓✓	S	S	22	✓	✓		
WELLS, Julie 881917	5. 5.63.												T/L/S	7			✓ 7/52	Respiratory infection Gross hydronephrosis

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
LOY, Mark 881969	7. 5.63.			✓								O	S	3	✓	✓		
MARSDEN, Jayne 882126	13. 5.63.								✓		✓✓	O	L/S	9	✓	✓		
BERRY, Joan 882128	13. 5.63.	✓	✓✓✓				✓		✓				L/S	50			✓ 4 yrs.	Renal failure
MOFFETT, Anna 882151	14. 5.63.			✓							✓	O	L	3	✓			
DUCKERING, Andrew 882210	15. 5.63.	✓			✓							S	L	6	✓	✓		Bright boy
BRAY, Jennifer 882265	18. 5.63.												T/L	2			✓ 2/52	Horseshoe kidney Ventriculitis Pneumonia
WESTERMAN, Diane 882337	20. 5.63.								✓		✓✓✓✓✓	S	L/S	18	✓	✓		
BARNES, Jennifer 882336	21. 5.63.												L/S	1			✓ 5/365	Pneumonia Pyocyaneus Septicaemia
GRAYSON, Jane 882356	22. 5.63.												L/S	1			✓ 1/365	No post mortem Intestinal perforation

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
KIRTON, Annette 882461	27. 5.63.	✓	✓								✓✓		T/L	24		✓		Low I.Q.
WAGSTAFF, Debbie 882531	28. 5.63.												T/L/S	3			✓ 3/52	Ventriculitis
WATHALL, Yvonne 882757	9. 6.63.												L	14			✓ 1 yr. at home	Chronic ventriculitis Bronchopneumonia Urinary tract infection Treatment abandoned at S.C.H. because of persistent meningitis after 14 weeks.
CROKER, Jane 882928	15. 6.63.												T/L/S	6			✓ 6/52	Ventriculitis No post mortem
LOFTUS, Sheila 883051	21.6. 63.												T/L	5			✓ 4/365	Infection - No post mortem
LEE, Paul 883071	23. 6.63.												L	13			✓ 3/12	Over active valve. Chest infection (head down nursing) No post mortem
GRITTON, Terry 883152	27. 6.63.	✓	✓✓		✓						✓✓✓✓ ✓✓✓ ✓✓✓	S	T/L/S	48				I.V.P.at 5 yrs.Hydronephrosis
CHAMBERS, Victoria 883766	22. 7.63.												T/L	1			✓ 4/365	Pneumonia
WALTERS, Mark 884045	4. 8.63.	✓	✓✓✓								✓✓✓✓	S	T/L/S	50		✓		Pyloric Stenosis

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
CATTERMOLE, Caroline 884299	16. 8.63.	✓	✓✓✓✓✓						✓		✓✓	S	L/S low	31	✓	✓		Hydronephrosis Long shunt in colon
OAKIN, Anne 884317	18. 8.63.	✓	✓	✓								O	S	8	Near normal	✓		Diastematomyelia
LAIGH, Glen Leslie 884548	28. 8.63.	✓	✓	✓							✓✓✓✓	O	L/S	12	✓	✓		Hemimyelocele
NORTON, Mark 884594	30. 8.63.	✓	✓✓	✓								O	S	8	✓	✓		
TROOP, Sophia 884634	2. 9.63.	✓	✓✓✓								✓✓✓✓✓✓	S	T/L/S	44	✓			Hydronephrosis at 5
HOUGHTON, Wendy 884757	8. 9.63.	✓	✓✓✓✓										L/S	28			✓ 8/12	Gross (L) side hydronephrosis and pyonephrosis. Post mortem. Died of chronic ventriculitis. Revisions included ventriculo-gastric shunt-Intussusception seen at P.M.
HILLAN, Mark 884735	12. 9.63.												T/L/S	1			✓ 4/365	Large ureters Post mortem done Infection
TAYLOR, Debbie 884784	9. 9.63.	✓	✓								✓✓	O	T/L	18	✓	✓		Apparent automatic emptying Hemimyelomeningocele

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
IMMS, Paula 884849	12. 9.63.												T/L	9			✓ 9/52	Bilateral hydronephrosis Horseshoe kidney Post Mortem. Ulcer and perforated lower oesophagus. Ventriculitis etc.
ROCKLEHURST, Kevin 884890	14. 9.63.	✓											T/L	5			✓ 4/12	Gastro-enteritis. Evidently at home. No post mortem
RADDOCK, Jacqueline 885045	23. 9.63.	✓	✓✓✓									S	L	15		✓		Some spasticity. Hemimyelocele. (L) leg only affected.
cELVANEY, Paul 885118	25. 9.63.												S	2			✓ 2/52	Diastematomyelia. Ventriculit Wound breakdown.
ILLIAMSON, Sharon 885176	29. 9.63.												L	10			✓ 10/52	Intraventricular haemorrhage
EE, Lynn 885222	4.10.63.	✓	✓✓✓	✓								O	T	16	✓	✓		Additional Lipoma of the cauda equina
OWLEY, Dawn 885346	7.10.63.												T/L/S				✓ 2/365	Probably intracranial haemorrhage
OWSER, Nigel 877834	13.10.63.												T/L/S	2			✓ 16/365	Ventriculitis
ORLER, Josephine 885577	15.10.63.												L				✓ 2/365	Ventriculitis. Post Mortem.

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				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
PEASGOOD, Philip 885644	19.10.63.	✓										S	T/L	5 + 3/12	✓	✓		Follow up elsewhere.
COOKE, Alistaire 885684	21.10.63.	✓	✓		✓						✓✓✓✓ ✓✓	O	T/L	21	✓	✓		Osteomyelitis
BROWN, Andrea 885761	23.10.63.												T/L/S	3			✓ 3/52	Septicaemia - wound infection
SWALLOW, Michael 885859	29.10.63.	✓		✓								O	T	7	✓	✓		
HOWLING, Dale 885763	24.10.63.										✓✓✓ ✓✓	S	L/S	10	✓	✓		
GARBETT, Mary 885764	24.10.63.												T/L/S	8			✓ 3/12	Ventriculitis
SANDERSON, Paula 885824	27.10.63.												T/L/S	-			✓ 2/365	Probably intracranial bleed
BLATHERWICK, Elaine 885923	31.10.63.										✓✓✓✓ ✓	S	S	6	✓	✓		Microcephaly
SPURR, Simon 885961	1.11.63.					✓					✓✓✓✓ ✓✓	S	S	4	✓	✓		
SAXTON, Julie 886015	5.11.63.										✓✓✓✓ ✓✓	O	T/L	22	✓	✓		Diastematomyelia Hemimyelocele

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
FROST, Christine 886296	14.11.63												T	2			✓ 2/52	Intracranial bleed Died of pneumonia
THOMSON, Donna 886365	19.11.63	✓	✓✓✓						✓			O	T/L	34				I.V.P. No dilatation but reflux. Ventriculitis treate
SMITH, Amanda 886465	26.11.63							✓	✓	✓	✓✓✓✓	S	L/S	32	✓			I.V.P. age 5. Non-function on (L) good (R)
BRAILSFORD, Peter 886435	23.11.63												L	5			✓ 6/12	Died at home? No post mortem
ALLS, Leonard 886697	5.12.63												T/L/S	-			✓ 1/365	Gastro-intestinal bleeding. Septicaemia Pneumonia.
MELLOR, Caroline 886768	8.12.63											O	S	21	✓	✓		Subarachnoid. Intraventricular and subdural bleeds. Grade I hydrocephalus. Minimal neural defect. Patulous anus. Sensory loss.
POTTS, Barbara 886861	12.12.63			✓								O	S	3				Normal child. Lesion almost completely covered with epithelium

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5'	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
HARDY, Angela 886860	12.12.63			✓								O	S	2				Normal child Lesion almost completely covered with epithelium
HARTLEY, Maria 887019	20.12.63	✓									✓✓✓✓ ✓✓	S	L	22	✓	✓		
SARGENT, Mark 887018	20.12.63												L	2			✓ 2/52	Good legs. Gastro-enteritis. No post mortem
McLENNON, Alison 887040	22.12.63												T/L/S	4			✓ 4/52	Single kidney Ventriculitis
JACKSON, Glenda 887064	24.12.63												L	15			✓ 3 yrs.	Hydronephrosis aged 2 Coroner's P.M. Gastro-enteritis + cone (Y) Re-exploration of back at 5/12, dermoid. Good legs at birth and until 5/12.
HALL, Darrell 887234	4. 1.64											O	S	3		✓		Normal
GREENWOOD, Katherine 887347	10. 1.64							✓					T/L/S	34			✓ 9/12	Gross (L) hydronephrosis at 2/52, (R) at 4/12. (L) loop ureterostomy. (R) uretero-appendicostomy. Death mainly renal.

NAME AND NUMBER	Date of closure	MICTURITION					URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
		Valve	Valve revision	Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
BRIDDON, Susan 887418	13. 1.64												L/S	6			✓ 5/12	Imperforate anus Cone. Died at home. Coroner's post mortem
PERRITON, Nigel 887525	18. 1.64	✓	✓			✓					✓✓	O	L/S	13	✓	✓		
WOODS, Kevin John 887458	15. 1.64	✓									✓✓✓	O	S	15	✓	✓		
WALASZEWSKI, Christopher 887529	17. 1.64												L/S	3			✓ 3/52	Ventriculitis Epidemic E.coli
McBRANE, Paul 887667	25. 1.64	✓	✓								✓✓✓ ✓✓	S	L/S	18	✓	✓		
HARTLEY, Tracey 887853	2. 2.64	✓	✓✓ ✓										T/L	26		✓		Frequent chest infections Low I.Q. and considered unsuitable for orthopaedic treatment
MELVILLE, Fiona 887906	4. 2.64	✓							✓			S	T/L	67	✓			Hydronephrosis uncontrolled
WHITING, Paul 888011	7. 2.64					✓					✓✓✓✓ ✓✓✓✓ ✓✓✓✓	O	L/S	12	✓	✓		—
LIVERSEDGE, Sheila 888046	10.2.64												T/L	12			✓ 8/12	Probable urinary failure. No post mortem

NAME AND NUMBER	Date of closure	MICTURITION					URINARY OPERATION					Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
		Valve	Valve revision	Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision									
THOMSON, William 888047	10. 2.64	✓		✓								✓✓	0	L/S	5	✓	✓		Hemimyelomeningocele
THOMAS, Susan 888063	10. 2.64													T/L	9			✓ 2/12	Bronchopneumonia
BROWN, Beverley 888096	12. 2.64								✓	✓		✓✓✓✓		L/S	21	✓			Uncontrolled hydronephrosis at 5
WILBY, David 888187	14. 2.64			✓									0	S	2	✓	✓		Anal stenosis
MOONEY, Paula 889029	25. 3.64	✓	✓					✓	✓	✓		✓✓	S	T/L	42				Hydronephrosis controlled at 5
WILSON, Robert 888343	22. 2.64	✓				✓						✓✓✓	S	L/S	8				Normal I.V.P. at 5
CLARK, Martin 888387	25. 2.64	✓	✓✓✓✓				✓✓					✓✓✓ ✓✓✓	S	T/L	50	✓			Early renal dilatation at 5
CHAPMAN, Hazel 888480	28. 2.64	✓	✓									✓✓✓✓ ✓✓✓	S	L/S	22	✓	✓		Given wheelchair at school
UNDERWOOD, Duncan 888503	29. 2.64	✓	✓✓✓✓				✓					✓✓✓ ✓✓✓	S	L/S	48	✓	✓		

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
ARCHER, Julie 888549	3. 3.64								✓	✓	✓✓✓	O	T/L	17	✓			Hydronephrosis and stone at 5
HARVEY, Denise 888571	4. 3.64												T/L	8			✓ 3/12	Urinary infection)Proteu Septicaemia)No P.M
TAYLOR, Darryl 888591	5. 3.64				✓						✓✓	S	L/S	9	✓	✓		Re-exploration of back - dermoid cyst
HOLLAND, Martin 889736	16. 6.64	✓	✓	✓								S	L	10	✓	✓		Almost skin covered but very small plaque.
GILYEAT, Caroline 888906	18. 3.64												S	2			✓ 2/52	Pyocyaneus ventriculitis This might have been a good child
BENSLEY, Jane 888926	19. 3.64	✓	✓	✓							✓✓✓	O	T/L	20	✓	✓		Hemimyelomeningocele
FINN, John 888973	22. 3.64	✓	✓		✓						✓✓✓	S	L	8	✓	✓		Hemimyelomeningocele
FOWLER, Brenda 889034	24. 3.64												T/L	3			✓ 3/52	Ventriculitis
ROBERTSON, Nigel 888972	1. 4.64												T/L/S	-			✓ 1/365	Died on table Lung hypoplasia

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
SHARP, Terry 889180	1. 4.64.	✓	✓✓								✓✓✓✓	S	T/L/S	29		✓		
WATHAM, Sally 889216	3. 4.64.	✓											T/L/S	10			✓ 4/12	Bronchopneumonia
PATRICK, Stephen 889255	6. 4.64.				✓						✓✓✓✓✓✓	S	L/S	11	✓	✓		Relatively small m.m. adequate bladder. Multip orthopaedic procedures. Walks in below knee calip
WALKER, John 889310	8. 4.64.	✓	✓	✓							✓✓	S	T	11		✓		Re-exploration of back - tethering. Spastic paraplegia
WALL, Stuart 889421	14.4. 64.	✓	✓✓					✓				O	L	27	✓			Solitary kidney Controlled hydronephrosis
DUCKMANTON, Anthony 889503	17. 4.64.	✓	✓✓✓								✓✓✓	S	L/S	18		✓		Low I.Q. Horseshoe kidney
BECK, Wendy 889578	21. 4.64.			✓								O	S	9		✓		
WILSON, Caroline 889693	25. 4.64.	✓	✓✓								✓✓✓✓ ✓✓✓	O	T/L/S	21	✓	✓		
BLOWER, Gary 889766	28. 4.64.			✓								O	T	5	✓	✓		Normal child though big head

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
SLATER, Christopher 890045	10. 5.64.			✓								O	S	2	✓	✓		Normal child. Very small plaque. Calf and gluteus maximus only.
ROWBOTTOM, Paul 890034	11. 5.64.	✓	✓										T/L/S	14			✓ 2 yrs.	Urinary tract normal thou double kidney. Brain stem haemorrhage
KILTY, John 890156	15. 5.64.	✓	✓✓✓								✓✓✓	S	T/L/S	46				I.V.P.at 5 years - early dilatation. Theco-peritoneal shunt at one stage. Excision of dermoi in back lesion
PRICE, Peter 890202	19. 5.64.	✓	✓								✓✓✓	O	T/L/S	56	✓	✓		
HODGSON, Joanne 890234	19. 5.64.												L/S	1			✓ 3/365	Probably intracranial haemorrhage + septicaemia
TAYLOR, James 890236	20. 5.64.	✓	✓		✓						✓✓	O	T/L/S	8	✓	✓		
GRAYSON, Neil 890235	20. 5.64.												T/L/S	2			✓ 2/52	Septicaemia
KIRKHAM, Lynn 890299	24. 5.64.				✓						✓✓	O	L/S	4	✓	✓		Diastematomyelia
PLEASANTS, Deborah 890402	28. 5.64.	✓	✓✓✓						✓		✓	S	S	19	✓	✓		

NAME AND NUMBER	Date of closure	MICTURITION				URINARY OPERATION					Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
		Valve	Valve revision	Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
WINFIELD, Mary 890399	28. 5.64.												T/L/S	1			✓ 2/365	Marked rib deformity. Died of inadequate respiratory function
WILLS, Kevin 890454	30. 5.64.	✓											L/S	5			✓ 5/52	Died at home? Infection
EAMES, Neil 890441	1. 6.64.			✓								0	L/S	6		✓		Quite extensive plaque but good leg movements at final examination. Flaccid feet
DAVEY, Linda 890544	3. 6.64.	✓											L/S	4			✓ 4/52	Septicaemia. Valve infecti
PEPPER, Ann 890618	5. 6.64.								✓		✓✓✓✓	0	L/S	12	✓	✓		
MUNTON, Dawn 891140	26. 6.64.												T/L/S	2			✓ 2/52	Sepsis. Ventriculitis
MORTIMER, Beverley 891367	3. 7.64.	✓	✓					✓	✓		✓✓	S	T/L	42	✓			I.V.P. Hydronephrosis but improved since diversi
SHORT, Janet 891402	6. 7.64.												T/L/S	16		✓	✓ 2 yrs.	Diastematomyelia Hemimyelomeningocele Re-exploration
BRAGAN, Ashley 891232	7. 7.64.	✓	✓		✓							0	L/S	8	✓	✓		

NAME AND NUMBER	Date of closure	Valve	Valve revision	MICTURITION			URINARY OPERATION				Orthopaedic operations	School Special (S) or (O)	Level of lesion	Weeks in hospital	Walking	Normal I.V.P. at 5	Died	NOTES
				Near normal	Acceptably dry	Appliance	Bladder outlet	Ureterostomy	Bowel conduit	Loop revision								
MULLINS, Anthony 891368	10. 7.64.											S	T/L/S	16	✓	✓		
PALLISTER, June 891513	12. 7.64.	✓	✓								✓✓	S	T/L	8		✓		
SNAZELL, Glen 891649	16. 7.64.												L/S	2			2/52	Laryngeal stridor
WATFORD, Kay 891848	25. 7.64.												T/L/S	7			✓	Pneumonia
CARTWRIGHT, Paul 891884	28. 7.64.										✓✓✓ ✓✓	S	L/S	7	✓	✓		
FAIRCLOUGH, Dawn 891907	29. 7.64.			✓							✓	O	L	8		✓		Hemimyelomeningocele
OVEREND, Jacqueline 892188	11. 8.64.								✓	✓		S	S	10				I.V.P. at 5, hydronephros controlled.

APPENDIX II

SHEFFIELD TELEGRAPH *μ.σ.π** 13.8.71

Doctor's new theory on spina bifida

By IAN MIDDLETON

Only children suffering from spina bifida who may look forward to a life without grave handicaps should be given the treatment to keep them alive, thinks a Sheffield doctor who is a world authority on the disorder.

He is Dr John Lorber, of Sheffield University Department of Child Health, who believes an accurate forecast of a child's minimum future handicap could be made in the first 24 hours after birth.

In a paper in Developmental Medicine and Child Neurology, Dr Lorber says: "If we wish to spare children and their families prolonged suffering and to give better attention to those who are more likely to benefit from total care, we may have to select suitable cases for intensive treatment and others for no treatment."

The overwhelming majority of untreated children would die within six months, according to figures quoted by Dr Lorber.

Restricted

He says a proportion of the 2,000 or so spina bifida babies born each year are "so severely handicapped at birth that no method of therapy could prevent major permanent multisystem handicaps, a short life expectancy, and a severely restricted quality of life."

Even after major surgery the more severely handicapped patients may suffer severe paralysis, mental retardation, incontinence, and other handicaps.

Dr Lorber analyses histories of 524 unselected spina bifida cases admitted since 1959 to Sheffield Children's Hospital, where for 12 years a policy of treating all patients offered has been followed.

He attempts to establish medical criteria which will show—in the first 24 hours after birth—the minimum degree of future handicap the child could expect if it survived.

These criteria could be used to select babies for treatment after birth.

Selection is already practised by some doctors, although there is a tendency towards the Sheffield policy of "treat all" said Dr Lorber last night.

He emphasised that it was for society to decide whether a general policy of selection should be operated. But if it was in favour, the selection should be done by consultants experienced in this field on purely medical grounds.

For this reason his paper "deliberately does not deal with the effect such a child has on family life or finances." He is concerned to make a "purely clinical assessment."

COMMENT — Page Six

Matter of life or death

MEDICAL science, which has concentrated so far on the unequivocal and unqualified saving of life (in theory and most practice anyway) has now reached the point where in many spheres the darker question of considering what sort of life is worth saving is becoming increasingly pressing.

Dr John Lorber, of the Department of Child Health at Sheffield University, has had the courage and the honesty to put that question in public.

So far most of the rest of us have been content to leave life and death to doctors and surgeons. In many ways they have taken over the function of the priest in a less formally Christian society.

In saying that babies with hopeless spina bifida should be well cared for, but allowed to die of natural causes, and only the more hopeful cases treated surgically, Dr Lorber is simply doing what he sees as his duty, putting the question to us which we have so far left willingly to him.

It is clear that in many medical fields, in care for the old and the chronically sick, the sort of decision that he now wants to be taken openly is already often taken in private. In other areas where Sheffield's "treat-all" approach to spina bifida children is not followed, selection for treatment must of necessity already exist unofficially.

As far as our layman's knowledge can tell, Dr Lorber's research, figures and technical knowledge are impeccable. His generous concern for the harrowing condition of many of his patients pulses through the dry medical surface of his article, and we honour him for it. But we still feel that he is assuming too much.

Subjective

His summary of his argument is that "objective data are presented which suggest that selection . . . can be made on a humanitarian basis." We cannot wholly agree. His assumptions are subjective. He is saying that in his opinion the sort of life his worst patients lead is not worth living.

The principle at stake is not a purely medical one. Dr Lorber's constant contact with such sufferers makes him highly qualified to give a personal opinion, but it can only be that. On a matter like this a consensus is needed — and society cannot, must not, allow an opinion to become a matter of automatic medical practice.

Dr Lorber, to his great credit, recognises that. It is why he published his article. He, and many of his colleagues, find the robes of a god too heavy for their human shoulders.

More vital

In the long term further knowledge may remove this particular question by making sure that babies are not born with spina bifida. Dr Lorber's initiative should not stop people from giving money for more research in his field. In fact it makes it even more vital.

In the short term such hope is of little comfort. It might be easier and perhaps kinder to shut our eyes. But as the power of medicine to prolong life pushes us ever nearer to the hazy border between life and death, we shall face many more such decisions. We ought to come to terms with that fact.

DAILY TELEGRAPH 13.8.71

When it is 'humane' to let a baby die

DAILY TELEGRAPH REPORTER

THE "humane thing" was to allow children suffering severely from spina bifida to die rather than treat them, Dr John Lorber of Sheffield University's Department of Child Health, said last night.

An authority on the disorder, Dr Lorber believes an accurate forecast of a child's minimum future handicap from it can be made in the first 24 hours after birth. About 2,000 cases occur annually in Britain.

He was commenting on a paper he had written on "Developmental Medicine and Child Neurology," in which he suggests that suitable cases for intensive treatment may have to be selected.

A detailed study of the initially less handicapped patients left no doubt that a higher survival rate and a better quality of survival would be possible by improved new techniques of treatment or by the better application of existing knowledge and facilities.

Quality of life

"It is also apparent," writes Dr Lorber, "that a proportion of patients are so severely handicapped at birth that no method of therapy could prevent major permanent multi-system handicaps, a short life expectancy and a severely restricted quality of life.

"If we wish to spare children and their families prolonged suffering and to give better attention to those who are more likely to benefit from total care, we may have to select suitable cases for treatment and others for no treatment.

"To do this it is necessary to correlate accurately observed data obtained on the first day of life with the subsequent progress of the children.

"Such a prospective analysis would give in any individual baby, with particular signs, probabilities of various severe handicaps and also of relatively minor handicap or of none."

If selection were possible the best time would be soon after birth before any therapeutic procedure was undertaken. At this stage the emotional involvement of parents and doctors was "less strong."

"If given the chance, many parents express the wish that their child should not be treated if it will mean permanent handicap with major disabilities, provided also that their child's life is not unduly prolonged."

Selection at a later stage was "emotionally more difficult."

Dr Lorber states that large numbers are so severly handicapped at birth that those who survive are bound to suffer from a combination of major physical defects. In addition, many would be retarded in spite of everything that could be done for them.

While it might be true that further advances could improve the outlook for many, the basic defect was usually so severe that the children would always be severely handicapped in spite of any foreseeable advances.

"Until quite recently far too many infants with spina bifida were allowed to die or deteriorate for want of expert care. The pendulum has now swung too far: there are now many with dreadful handicaps who a short time ago would have died."

SHEFFIELD TELEGRAPH 13.8.71

LETTERS to the EDITOR

Spina bifida: no change of policy

SIR, — The consultant surgeons responsible for the treatment of spina bifida patients at the Children's Hospital in Sheffield wish to express their views in relation to your quotations from Dr Lorber's recent paper on the subject. The lay Press is not the place to discuss the details of a medical article from which the quotations were taken, but we would agree with the principal results of this analysis of records, namely that the infants who are severely affected at birth are severely handicapped later on.

It is only fair to Dr Lorber to make it clear that these babies were not his responsibility when admitted to the hospital and few, if any of them, were seen by him before operation.

From our own observation of these babies on the first day of life and from their subsequent progress, several important points emerge.

1. The primary purpose of treatment is to improve the quality of life of the child.

2. The operation on the back of the new born baby is not intended to be, nor is it, a "life-saving" procedure.

3. We have seen considerable numbers of untreated babies survive, perhaps for months or even years, more disabled than they need to be and in chronic ill health: in fact, most of our work in the past was concerned with untreated older children who had survived with severe deformity and disability.

4. Many severely affected babies can be helped considerably by surgery, not only to prevent disabling complications but to reduce the handicaps that are already present.

5. If our results fall below our hopes and expectations, we regard this as a challenge to further research to improve techniques — rather than an indication to abandon any attempt at treatment, in the hope (not always fulfilled) that the child may die soon from so-called "natural causes".

We would like to correct any impression which might have been gained from the article that there has been any fundamental change in policy at the Sheffield Children's Hospital in the treatment of spina bifida, treatment which is almost entirely a surgical problem.

We adhere to our aim to reduce the handicap to the minimum and to develop the child's abilities to the maximum and, in addition, we are concerned that the families of spina bifida children should have the support of knowing that the doctors are doing all they can to help their children.

R B Zachary, FRCS
W J W Sharrard, FRCS
James Lister, FRCS

Children's Hospital,
Western Bank,
Sheffield.

25.8.71

SCOTTISH DAILY EXPRESS

BABIES WITHOUT HOPE...

ABOUT 15 BABIES brought into an Edinburgh hospital every year suffering from severe spina bifida are not being given operations to help keep them alive.

This was revealed yesterday by Dr. Gordon Stark, a consultant physician at the Royal Hospital for Sick Children.

And he has added fresh fuel to the nationwide controversy which flared earlier this month when a Sheffield doctor, after following up 500 cases, also claimed that severely affected babies should not be treated.

'Scepticism'

Dr. Stark said at the hospital yesterday: "We have pursued a selective policy since the unit was opened five or six years ago, not to give the operation to every patient. This policy has been vindicated by the Sheffield results."

In an article in the Archives of Disease in Childhood, Dr. Stark stated: "With traditional Scottish scepticism, the Royal Hospital for Sick Children is among a minority of units in which it is felt that some of these infants are so severely handicapped, and so unlikely to survive, that operation can not be justified.

"In most large centres in the United Kingdom, on the other hand, it is standard practice for every infant with a myelomeningocele (the technical name for the condition) to be subjected to early operation, irrespective of his condition or prospects."

Before recent advances in surgery, many of these babies were registered as "stillborn."

But whatever the ethical position, Dr. Stark added, every one of these infants deserves an opportunity for "full assessment" immediately after birth, to see if an operation should be given.

The Royal Sick Children's Hospital in Edinburgh serves a population of about 1,000,000, and admits 35 new cases of spina bifida a year.

Between 40 and 45 per cent are not operated on, according to Dr. Stark.

"It is true without a crystal ball, we can not tell which child's burden of disability will be increased."

But assessment soon after birth could indicate the least amount of disability the infant might later be forced to suffer if an operation was performed.

"Miracles may happen, but they are rare in this field," Dr. Stark added.

He quotes Dr. John Lorber, of the Department of Child Health at Sheffield University, who has already said that both survival and quality of later life are badly endangered if certain signs are found.

"It is likely that the disappointing Sheffield results will lead to general reappraisal of the policy of routine early operation," Dr. Stark pointed out.

by

ALISTAIR CAMPSIE

"Miracles may happen, but they are rare in this field"

—DR. GORDON STARK

children's surgeon said in Glasgow yesterday: "Really, to prolong life in this condition can be quite ridiculous.

"But this is dangerous stuff to speak about. It is too emotional for most people involved to discuss rationally."

Dr. Stark said at the hospital yesterday: "Most of these children die if not operated on, but survival times are very variable.

"Some live for a few days or a few weeks, and some can live on indefinitely. They are kept as comfortable as possible in the wards."

Miss Mary Oughtred, general secretary of the Spina Bifida Association said in London yesterday: "We exist to give every help and support to those with spina bifida and their families, and to raise funds

"But treatment and who should be treated are medical matters, between doctors and the parents of the child. We cannot comment on this."

A Church of Scotland spokesman said in Edinburgh: "In 1968 the Church and Nation Committee reported on the ethics involved in modern medicine, notably prolonged life.

"They said they were fully aware of the burden of responsibility new skills and techniques laid upon the medical profession.

"They said that every patient was individual and must be treated as such. Each case must be treated on its merits, and every doctor must take his own decision."

A leading Catholic theologian commented: "This is a medical matter and one which can only be solved by doctors."

RESEARCH NOW!

THE report that certain babies suffering from spina bifida — a disabling and almost invariably fatal malformation— do not receive surgical treatment in some Scottish hospitals must trouble parents and the public.

It is further evidence — all the more tragic where young children are concerned — of the grave dilemma that so often faces doctors and surgeons.

We know that there are "hopeless" cases. People beyond the cure of surgery. There are thousands of people with bad hearts who would never be considered for a transplant and who probably would not want one.

To inflict further useless suffering in such cases is just as bad as neglecting them. To say nothing of the possibly vain raising of hopes.

The one positive action we can take is to press all the harder for the research that will produce a cure. Then patients, parents and surgeons can be spared the terrible decision now thrust upon them.

Scottish Daily Express 25.8.71

APPENDIX III

Electrolyte Absorption From Isolated Bowel Segments Used as Urinary Conduits

By R. C. M. Cook, C. I. Franks, J. Lister, J. Pratt and R. B. Zachary

Reprint from February 1968 issue of Journal of Pediatric Surgery

Electrolyte Absorption From Isolated Bowel Segments Used as Urinary Conduits

By R. C. M. Cook, C. I. Franks, J. Lister, J. Pratt and R. B. Zachary

THE OPERATION of uretero-sigmoidostomy has now been almost entirely abandoned because of the very high incidence of pyelonephritis, acidosis and hyperperchloremia. In its place diversion of the urine through an isolated segment of small intestine has become standard practice, first for adults after pelvic evisceration for malignant disease[1] and more recently[2,3] for children with urinary incontinence. There is a remarkable lack of exact information on the absorptive functions of the intestinal conduit used in this way.

There are a number of clinical reports[7-9] of acidosis in adults after this method of diversion and some adult and animal experimental work has been reported,[10-12] but we have been unable to trace any work relating to young children, either specifically to the normal absorptive functions of the terminal ileum or to the long term function of their urinary conduits.[13]

There has been a number of children in this unit in whom diversion of the urine through an ileal conduit was immediately followed by gross electrolyte upsets—plasma bicarbonate levels fell and the plasma chloride rose considerably. After a month or two of therapeutic adjustment of their serum electrolytes, the pattern returned to normal and has been maintained as such. These were all children who had some degree of impairment of renal function preoperatively.

There are a number of questions that immediately spring to mind when considering the intestinal urinary conduit. To what extent does it reabsorb the urinary constituents? Does the passage of urine over the mucosa for a long time alter its absorptive functions? Does its motility favor retention or expulsion of the contents, or does it only mix them? What bacteriological flora, if any, must be considered normal in a urinary ileostomy?

We have particularly been concerned with the first two questions, although we allow that the third has an obvious bearing[14] and that the fourth is of vital importance to the continuing efficiency of the entire urinary system. The work reported here is a preliminary survey to see how some of these problems might be investigated.

The simple closed loop of intestine (Fig. 1A) has been studied in some detail in various centers,[11,15,16] but the solutions used have been of electrolytes in concentrations comparable to plasma, but quite unlike the rather extraordinary composition of urine.

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The work of R. C. M. Cook and J. Pratt was supported by a grant from "Action for the Crippled Child."

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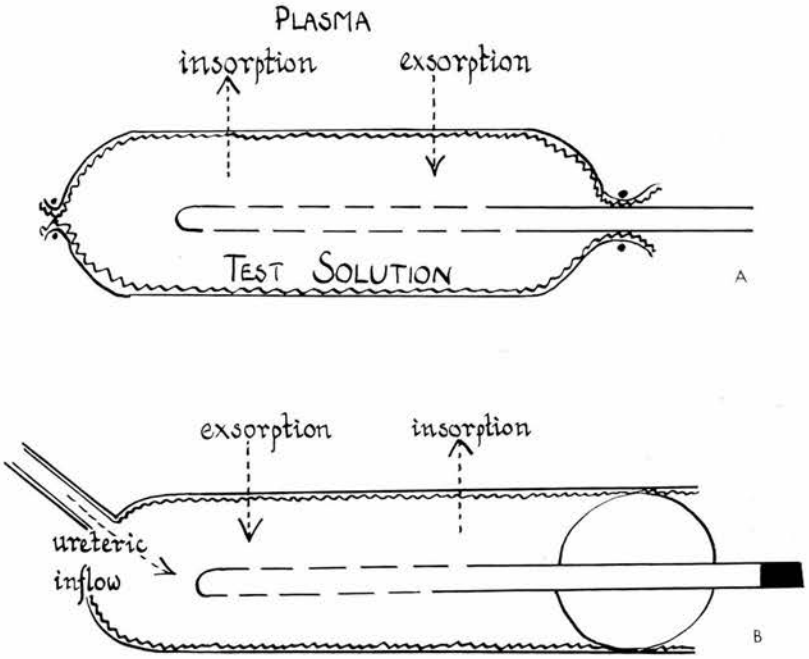

Fig. 1.—A, Diagram of simple isolated loop of intestine. **B,** Diagram of experimental arrangement of ileal conduit.

In the ileal conduit in a child (Fig. 1B) there are four flows[17] with which we are concerned: ureteric inflow and exsorption and stomal outflow and insorption. Insorption and exsorption are the fluxes across the wall of the conduit and the difference between them will be the net absorption or secretion. Stomal outflow can be directly measured and insorption calculated from the loss of constituent electrolytes from the loop, but we know of no way of distinguishing ureteric inflow from exsorption.

Urine samples collected from different levels within the ileal conduit through a triple-lumen catheter and samples collected before and after obstructing the loop for a period were analyzed. With these simple tests it was only possible

Table 1.—*Fluxes of Sodium*°

Case No.	Sodium Inflow via Ureters Prior to Test (Microequivalents per Minute)	Mean Sodium Concentration in Conduit Milliequivalents per Liter)	Total Inflow of Sodium Into Conduit (k_1) (Microequivalents per Liter)	Insorption (k_2) (Microequivalents per Minute)	$\frac{k_2}{k_1} \times 100$
1	79	176	94	26	28
10a	50	54	55	19	34
10b	70	78	78	10	13
11a	68	78	178	59	33
11b	76	88	144	68	47
12	40	54	63	26	41
13	60	88	107	35	33

°Calculation is similar to the method described by Duthie and Atwell[15] and is further elaborated in the Mathematical Appendix.

Table 2.—*Fluxes of Potassium*°

Case No.	Potassium Inflow via Ureters Prior to Test (Microequivalents per Minute)	Mean Potassium Concentration in Conduit (Milliequivalents per Liter)	Total Inflow of Potassium Into Conduit (k_1) (Microequivalents per Minute)	Insorption (k_2) (Microequivalents per Minute)	$\frac{k_1}{k_2} \times 100$
2	16	15	12	5	37
3a	18	37	47	27	57
3b	18	20	23	16	70
4a	44	85	71	20	28
4b	44	61	36	14	39
5a	43	50	48	12	25
5b	43	46	53	12	23
10a	53	54	57	12	21
10b	53	48	46	5	11
11a	32	37	88	23	26
11b	32	37	68	38	56
12	50	72	84	45	53
13	40	61	49	17	35

°Calculation is similar to the method described by Duthie and Atwell[15] and is further elaborated in the Mathematical Appendix.

to measure concentration changes without knowing how much they were due to water as opposed to electrolyte fluxes. They did suggest that potassium and chloride concentrations in the conduit fell and that sodium increased.

Na^{24}, K^{42}, Br^{82} (4 microcuries each) and tritium (20 microcuries) were used in label urine. In principle the method has been to remove the residual urine, to collect a sample from the base of the loop, as the nearest possible equivalent to ureteric urine, and then to put labeled urine into the conduit (Fig. 1B). It is kept obstructed for a short period and then emptied, washed out and isotope and stable electrolyte analysis done. (Total body irradiation 30 millirad., mucosal dose 200 millirad.) With outflow stopped, any changes should be due to ureteric inflow and exsorption. Tables 1–4 show the insorption in microequivalents per minute. In each table it is apparent that there is a very wide

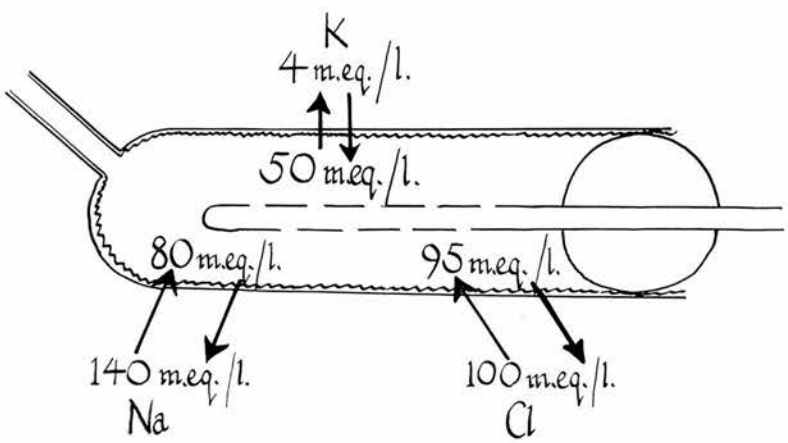

Fig. 2.—Typical concentration gradients across the wall of an ileal conduit.

Table 3.—*Fluxes of Chloride (Bromide)*°

Case No.	Chloride Inflow via Ureters Prior to Test (Microequivalents per Minute)	Mean Chloride Concentration in Conduit (Milliequivalents per Liter)	Total Inflow of Chloride Into Conduit (k_1) (Microequivalents per Minute)	Insorption (k_2) (Microequivalents per Minute)	$\frac{k_1}{k_2} \times 100$
6	60	95	61	20	33
8	40	120	51	19	37
9	23	157	27	10	37
10a	60	65	85	36	42
10b	60	68	77	28	36
12	50	68	87	43	44
13	60	85	98	55	56

°Calculation is similar to the method described by Duthie and Atwell[15] and is further elaborated in the Mathematical Appendix.

range of flow from the kidneys and of the mean concentration of each electrolyte within the conduit. The total inflow (k_1) and the insorption (k_2) have been calculated and the final columns show insorption (that is, the flux back into the plasma from the lumen) as a percentage of the total inflow. For sodium this ranged from 13 to 47 percent, for potassium between 11 and 70 per cent and for chloride (or bromide) between 33 and 56 per cent. Somewhere between 0.5 and 1.5 ml./min. of water was insorbed, but the total flow into the conduit was also considerably different from the probable ureteric inflow, and so considerable exchanges appear to be taking place.

If we consider the concentration differences on the two sides of the wall of the conduit (Fig. 2), exsorption of potassium would probably be very small compared with our figures for insorption. If this is so, then our figures in the

Table 4.—*Fluxes of Water*°

Case No.	Urine Inflow (ml./min.)	Mean Volume of Conduit (ml.)	Total Inflow of Water Into Conduit (k_1) (ml./min.)	Insorption (k_2) (ml./min.)
1	0.5	7.5	1.0	0.6
2	1.1	12	1.5	1.0
3a	0.5	32	2.9	3.1
3b	0.5	30	2.2	2.4
4a	0.4	19	1.5	0.8
4b	0.4	10	1.3	0.9
5a	1.0	20	2.0	1.4
5b	1.0	39	2.9	2.4
7	0.3	20	1.6	0.8
8	0.3	24	1.1	0.7
9	0.2	12	0.7	0.6
10a	1.0	32	2.3	2.4
10b	1.0	34	2.4	1.9
11a	0.8	32	4.0	2.8
11b	0.8	34	4.0	3.3
12	0.7	25	2.0	1.3
13	0.7	29	2.4	1.9

°Calculation is similar to the method described by Duthie and Atwell[15] and is further elaborated in the Mathematical Appendix.

Table 5.—*Possible Extra Work Load on Kidneys of a 20 Kg. Child of 5 Years, With Urine Diverted Through an Ileal Conduit*

	Possible Daily Renal Excretion Rates[17] (Milliequivalents per 24 Hours)	Lowest Measured Insorption (k_2) (Microequivalents per Minute)	(Milliequivalents per 24 Hours)	"Extra Load on Kidneys"
Na	40	10	14.4	36%
K	35	12	17.3	50%
Cl	35	10	14.4	41%

potassium table (Table 2) represent something approaching the net absorption of potassium. For sodium and chloride, exsorption may well be more significant and so our figures will be greater than the actual net absorption. So if we take some approximate daily total renal excretion rates (Table 5) for a 5 year old 20 Kg. child, and compare these with the lowest experimentally found insorption, there may be reabsorption of 35 per cent of the sodium excreted each day, 50 per cent of the potassium and some 40 per cent of the chloride. For sodium, and perhaps for chloride, these figures may well be much smaller in practice, but it seems clear that reabsorption is potentially of a considerable degree.

There is of course a very wide variation in the measured insorptions, which on the small number of tests we have done we have been unable clearly to relate to factors such as the size of the conduit, the composition of the ureteric urine, the duration of obstruction or the age of the conduit. Some in vitro work we are doing suggests that the age of the conduit may be of some importance.

Conclusions

It has always been assumed that there was some reabsorption from an ileal conduit, but the actual measurement of this reabsorption, though difficult and showing wide variation, clearly demonstrates that it adds a considerable extra work load to the kidneys. The clinical inference is that ileal conduits should be avoided in patients who do not have a good reserve of renal function.

SUMMARIO IN INTERLINGUA

On ha semper suspicite que un conducto ileal resulta in un considerabile reabsorption. Le mesuration de iste reabsorption es difficile, e illo demonstra considerabile variationes. Nonobstante, le valores assi obtenite prova que le reabsorption ab un conducto ileal adde un carga considerabile al travalio del renes. Le conclusion a derivar ab le puncto de vista clinic es que conductos ileal deberea esser evitate in subjectos in que le reservas functional del renes non es bon.

ACKNOWLEDGMENTS

We would like to express our appreciation to Dr. R. J. Levin of the Department of Physiology for his help and interest and for the use of his laboratory, to Dr. H. Miller of the Regional Medical Physics Department, Sheffield No. 3 Hospital Management Committee, for his help and the use of the facilities of his department, and to Dr. Jordan and the staff of the Biochemistry Department of the Children's Hospital, who were responsible for the stable electrolyte analyses.

Mathematical Appendix

Notation

t = time (min.).
S_0 = total amount of stable substance in loop at time $t = 0$ (ml. or mEq.).
S_t = total amount of stable substance in loop at time t.
R = total amount of tracer in loop.
a_0 = specific activity of tracer in loop at time $t = 0$ (e.g., counts/sec./ml.).
a_t = specific activity of tracer in loop at time t.
k_1 = rate of inflow of stable substance into the loop, i.e., this includes urine flow and exsorption.
k_2 = rate of outflow of stable substance from the loop, i.e., insorption.

Calculations

Assume: (i) k_1, k_2 are constant with respect to time; (ii) the loop is in contact with an effectively infinite reservoir, e.g., for tritiated water this is the total body water volume and for potassium it is the total exchangeable body; (iii) mixing is perfect; this is very difficult to obtain in practice.

Then $R = aS$.

Differentiating with respect to time:

$$\frac{dR}{dt} = \frac{d}{dt}(aS)$$

$$= -k_2 a.$$

Now

$$S_t = S_0 + (k_1 - k_2)t. \tag{1}$$

Hence

$$\frac{dR}{dt} = \frac{d}{dt}\left\{ a\ [S_0 + (k_1 - k_2)t] \right\}$$

$$= \frac{da}{dt}\left\{ S_0 + (k_1 - k_2)t \right\} + a\,(k_1 - k_2)$$

$$= -k_2 a$$

or

$$\frac{da}{a} = \frac{-k_1 dt}{\left\{ S_0 + (k_1 - k_2)t \right\}}$$

Integrating, we have:

$$\ln a = \frac{-k_1}{k_1 - k_2} \ln \left\{ S_0 + (k_1 - k_2)t \right\} + \text{const.}$$

$$\text{Hence, } \ln \left\{ \frac{a_t}{a_0} \right\} = \frac{-k_1}{k_1 - k_2} \ln \left\{ \frac{S_t}{S_0} \right\}. \tag{2}$$

Solving equations (1) and (2), we have:

$$k_1 = \frac{\ln \left\{ \frac{a_t}{a_0} \right\}}{\ln \left\{ \frac{S_t}{S_0} \right\}} \times \frac{(S_t - S_0)}{t}$$

$$\text{and } k_2 = \frac{(S_t - S_0)}{t} \times \left\{ \frac{\ln \left\{ \frac{a_t}{a_0} \right\}}{\ln \left\{ \frac{S_t}{S_0} \right\}} - 1 \right\}.$$

Hence k_1 and k_2 can be found if a_0, a_t, S_0, S_t and t are known. S_0 includes the residual from isotope dilution at the beginning of the test and S_t includes the volume or quantity of electrolyte equivalent to the washings at the end of the experiment.

If k_1 and k_2 are low compared with the total quantity of stable substance in the loop, then it is reasonable to use the following simple equations:

$$k_2 = \frac{\text{loss of radioactivity}}{\text{mean radioactivity in loop}} \times \frac{\text{mean stable content of loop}}{\text{time } (t)}$$

and then

$$k_1 = k_2 + \frac{(S_t - S_0)}{t}.$$

This simple treatment is adequate with potassium but not with water, where the diffusion rate is high.

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Pressure Variations in Intestinal Loops used for Urinary Diversion

E. PEKAROVIC*, A. ROBINSON, J. LISTER and R. B. ZACHARY

Urinary diversion by implantation of the ureters into an isolated segment of ileum or sigmoid colon has found a wide application in the management of neurogenic bladder. There is no doubt that this procedure has proved its merit in preventing further dilatation of the upper renal tract, and in making these patients more socially acceptable.

After some experience with this operation the operative mortality is low. There are, however, quite significant late complications involving the abdominal stoma, such as stenosis, prolapse, epithelial dysplasia of the mucosa and bleeding. In addition there may be stone formation and also difficulties in fitting a suitable appliance. One or more of these complications is said to occur in 15–43 per cent of cases (Straffon *et al.* 1963, Rickham 1964, Eckstein 1965, Nash 1967).

Apart from these complications, further deterioration of the upper urinary tract may occur. Intravenous pyelograms performed after this operation may show progressive dilatation of the upper renal tract, found by some workers in as many as 30 per cent of their cases (Bricker 1952, Bricker *et al.* 1954). Chronic urinary infection may contribute to the dilatation, but stenosis at the stoma is a more likely cause. However, in some of these failures there is no evidence of stenosis at the level of the stoma in the abdominal wall, nor at the level of the uretero-intestinal junction.

One of us (Zachary 1967), suggested that at least in some of the cases of urinary diversion through an intestinal loop, the kidneys may be working against a high pressure system. It was felt that this question needed further investigation and various possibilities were considered.

Methods

In recent years many sophisticated electronic devices have been used for the investigation of pressure variations in the various systems of the body. Most of these can be applied to the urinary system. Pressure transducers may be used for the simultaneous recording of several urodynamic factors. After some experience with these techniques of bladder pressure measurement we decided to use similar techniques to find out whether increased pressure in intestinal loops could be a contributing factor in causing progressive hydronephrosis after urinary diversion.

We first attempted to measure the pressures by inserting a large catheter into the ileal loop and connecting it to the transducer and recorder, but the catheter was pushed out by the peristalsis of the loop. It is possible, too, that a large catheter might block the stoma and reduce the free outflow of urine, so increasing the pressure inside the loop. The use of a fine catheter was also unsuccessful, since this became blocked with mucus.

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*In receipt of a grant from 'Action for the Crippled Child'.

Another possibility is the use of a small balloon at the end of a fine catheter to measure the pressure indirectly. The disadvantages are that the balloon is not registering true intraluminal pressure, it is subject to squeezing by peristaltic waves of the intestinal wall and it is easily pushed out of the loop by its peristalsis.

A pressure-sensitive radio pill has already been used successfully in the female bladder by Shelley (1965) and in the human gut by Watson *et al.* (1966). The pill transmits information by means of radio waves to a receiver, the frequency being determined by the pressure surrounding the capsule. However, the pills available at the moment have a diameter of 9 mm., which makes them too large to introduce into loops with an average or narrow stoma.

Another possibility is a micromanometer designed by Holm (1966) for measuring intravesical pressure. Measuring 26 mm. long and 3 mm. in diameter, it would be small enough for any stoma. It is a mechanical device with a radio-opaque marker, so the pressure can be determined radiographically.

As neither this micromanometer, nor a suitable small radio pill is available to us at present, we have used a small transducer placed inside the loop. This transducer is approximately 3 mm. long and 3 mm. in diameter and is attached to the end of a number 6 French gauge catheter through which three fine wires pass to the amplifier and recorder. The air within the catheter provides free communication from the transducer to the outside, so that the transducer always measures the pressure relative to the atmosphere.

This is an advantage over the usual technique in which the transducer is placed outside the body and needs to be corrected for hydrostatic pressure, if it is not at the same level as the end of the catheter.

However, the transducer is sensitive to variations in temperature, and needs to be calibrated at 38°C. before insertion. The scale of our recorder is calibrated in cm. of water.

In order to increase excretion by kidneys and to promote full action in the loop, we give the patients plenty to drink before and during the investigation. Since the size of the catheter is small, we do not think it seriously interferes with the function of the stoma.

Observations

So far in our studies we have observed two different types of recordings. In one group of patients the line remains quite steady, showing the pressure around 10 cm. H_2O. This pressure hardly changes at all, even if patients increase their intra-abdominal pressure by straining, coughing, changing position, or by any muscular activity. (Figs. 1 and 2).

Fig. 1.

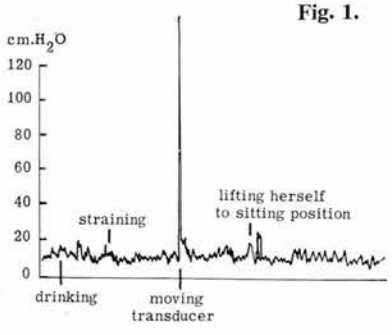
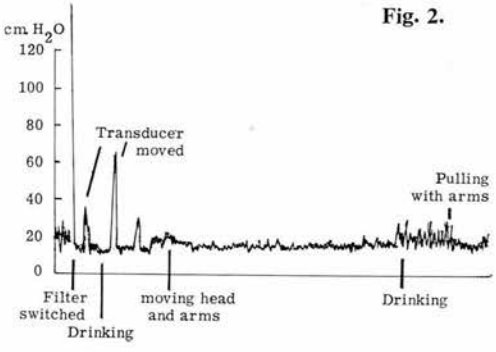


Fig. 2.



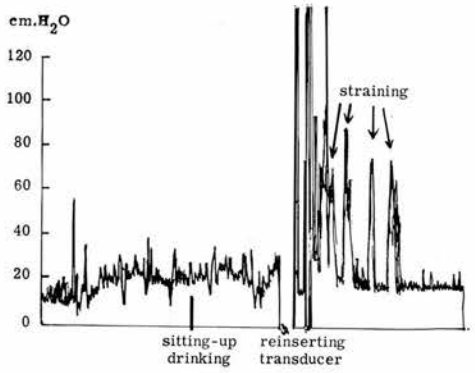


Fig. 3.

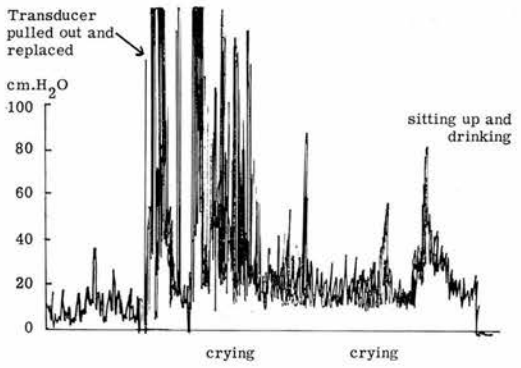


Fig. 4.

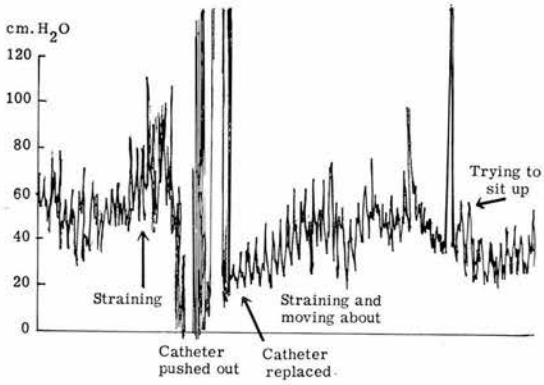


Fig. 5.

In the second group of patients the trace usually shows a higher basic pressure which is around 30 cm. H_2O, with sharp elevations reaching 60–80 cm. H_2O or occasionally even as high as 100–140 cm. H_2O when the patient is moving, straining, coughing, or with manual pressure. (Figs. 3, 4 and 5).

Before we can assess the value of these observations, we must consider the normal variations of intestinal pressure and the pressures normally found within the renal pelvis.

Using pressure transducers, Kiil (1957) found the resting pressures in the normal renal pelvis to be about 10 mm. Hg, with fluctuations of not more than 3–4 mm. Hg. This would be equivalent to about 15–20 cm. H_2O. The pressure within a hydronephrotic renal pelvis was not essentially different from that of the normal.

We had the opportunity to measure the pressure in the renal pelvis through a ureterostomy on three occasions using our small transducer. The pressures were 10, 12, and 20 cm. H_2O respectively, which would confirm the findings of Kiil.

The pressures measured within intestinal loops by Connell and Rowlands (1960) showed levels of 0–10 cm. H_2O in both ileum and pelvic colon rising only very occasionally up to 25–40 cm. H_2O. This would mean that under the normal condition the ureters would not be draining into a particularly high pressure system.

Moreover, taking into consideration the hydrostatic factor in the erect position, the pressure gradient from kidney to intestinal loop would be even greater.

The findings in the first group of our measurements showing the pressures of 5–25 cm. H_2O are within this normal range. If the intraluminal pressure of the intestinal loop was

the sole cause of dilatation of the upper renal tract, one would not expect deterioration in this group of cases.

On the other hand, deterioration of the upper urinary tract could be expected when the pressures within intestinal loops are high, as found in our second group of patients.

Results

Intraluminal pressure studies have been undertaken in 20 patients. These were not a completely unselected group, but included some patients who were examined specifically because of deterioration, but none had stomal stenosis.

In 9 cases the intraluminal pressures of the intestinal loop were found to be high (Table I).

In 2 of these 9 patients it was not possible to say whether there had been deterioration of the upper renal tract or not, since the pelvis and calyces were already grossly dilated before the ileal loop was constructed. However, in neither of these two cases was there any improvement. In the remaining 7 cases with a high pressure within the lumen, there was deterioration of the upper renal tract.

TABLE I

High Pressure Group

Case	*Pressure (cm.H_2O)*		*Loop Position*
	Resting	*Straining*	
1	5–15	50	Extraperitoneal
2	5–10	60	,,
3	10–30	60	,,
4	20–30	80	,,
5	20–40	100 (140)	,,
6	10–30	100 (130)	,,
7	20–30	60	,,
8	10–15	50	,,
9	15–25	90	,,

TABLE II

Low Pressure Group

Case	*Pressure (cm.H_2O)*		*Loop Position*
	Resting	*Straining*	
10	18–20	30	Extraperitoneal
11	5–15	32	,,
12	15–20	35	,,
13	5–10	20	,,
14	5–10	25	,,
15	15–20	30	Intraperitoneal
16	5–15	20	,,
17	10–12	15	,,
18	5–10	20	,,
19	5–10	25	,,
20	5–10	30	,,

A low intraluminal pressure was found in 11 patients, all except one having a good upper renal tract with no evidence of progressive dilatation (Table II). The only exception to this group (Case No. 10) has a long loop with considerable residual urine and persistent infection, and in her case considerable deterioration of the upper renal tract has occurred.

We have not yet been able to arrange for a re-examination of this child to see whether the emptying of the loop by a nurse immediately before investigation may not have been responsible for a lower intraluminal pressure than she normally has. This false impression of a lower intraluminal pressure has been demonstrated in another child.

Discussion

These investigations support the view that an intestinal loop with a low intraluminal pressure is likely to function well with no deterioration, whereas a loop with a high intraluminal pressure is likely to show progressive hydronephrosis.

Possible explanations of these findings are: (*a*) some technical error in the construction of the ileal conduit, (*b*) it might simply be the time factor, or (*c*) there might be some intrinsic factor causing a high intraluminal pressure in some patients and not in others.

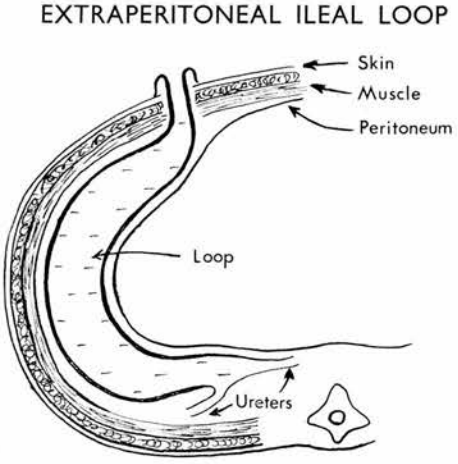


Fig. 6.

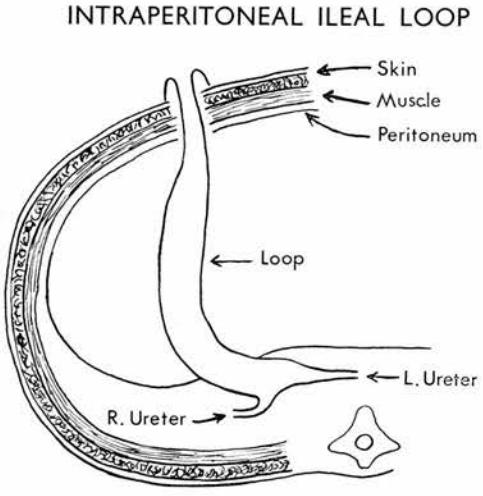


Fig. 7.

(*a*) *Technical error*

Prior to eighteen months ago the ileal loops were always constructed extra-peritoneally (Fig. 6), but more recently only the proximal part of the loop with the uretero-ileal anastomoses has been placed in an extra-peritoneal position, the remaining short conduit passing straight across the peritoneal cavity (Fig. 7). It is possible that such an extra-peritoneal position might lead to kinking of the loop, with some element of obstruction, but without any evidence of stenosis at the stoma itself.

In the high pressures group all 9 patients had their loop situated extra-peritoneally. In the low pressures group 5 out of 11 loops were situated extra-peritoneally.

However, kinking in a rather long loop cannot be the sole explanation because we have evidence of several loops which are short and with no evidence of kinking on a radiological study of the loop, but in which high pressures were found. A further possibility is that the position between the peritoneum and the abdominal musculature is one which is somewhat more likely to give high pressure than a loop within the abdominal cavity.

(*b*) *Time factor*

Those children who have had an ileal loop for a long time might be more liable to high intraluminal pressures than the recently constructed ones, although the exact explanation of such a deterioration in time is not at once apparent.

In this series, neither the extra-peritoneal position nor the time factor offers a complete explanation of high intraluminal pressure, since low pressures were found in some extra-peritoneal loops which have been present for longer than 18 months.

(*c*) *Intrinsic factor*

A third possibility of an intrinsic factor peculiar to some patients which leads to a high intraluminal pressure cannot be excluded.

This factor was suspected in a patient with a ureterosigmoidostomy in continuity several years ago. He was found to have progressive dilatation of the upper renal tract. At operation there was no evidence of stenosis at the uretero-sigmoid anastomosis, and needle aspiration of both ureters showed no infection. Intrarectal pressure was 90 cm. of water and this

high intraluminal pressure seemed to have been the cause of his deterioration. It does not seem impossible that there should be similar extreme variations in intraluminal pressures in the ileum.

It is important to detect poorly functioning intestinal urinary conduit before it has led to progressive dilatation of the upper renal tract. We believe that measurement of intraluminal pressures may give early warning of such poor function, whatever may be its mechanism.

SUMMARY

Intraluminal pressures have been studied in intestinal urinary conduits in 20 children with neurogenic bladders. High pressures are associated with progressive hydronephrosis.

RÉSUMÉ

Effets des variations de pression dans les anses intestinales utilisées pour la dérivation urinaire

On a étudié les pressions dans la lumière intestinale après dérivation urinaire, chez 20 enfants présentant des troubles neurologiques de la vessie. La présence d'une haute pression entraine une hydronéphrose progressive.

ZUSAMMENFASSUNG

Druckänderungen in den zur Umleitung des Urins verwendeten intestinalen Urinkanälen

Der intraluminale Druck in intestinalen Urin-Kanälen wurde bei 20 Kindern mit neurogener Blasenstörung registriert. Hoher Druck findet sich bei Kindern mit progressiver Hydronephrose.

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INDICATIONS FOR MANUAL EXPRESSION OF THE NEUROGENIC BLADDER IN CHILDREN

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In lesions involving the nerve supply to the lower limbs the bladder is nearly always involved, and even when there is no paralysis of the legs in a low lesion, the sacral outflow may be affected, leading to a neurogenic bladder. Hence lack of full control of the bladder is the most common major complication of myelomeningocele. Indeed, in a short series studied some years ago, out of 102 cases of myelomeningocele there were only two with apparently perfectly normal bladder control.

This figure contrasts with those of Carlson (1966) and Eckstein (1968) who reported a high proportion of continence in spina bifida children (20 to 40 per cent.), but these authors included in their series less serious types of spina bifida in which the spinal cord was not involved. Our findings agree more with the 11 per cent. incidence quoted by Cooper (1968) and the 2 normal bladders out of 111 by Smith (1965).

Full control of bladder function is therefore probably exceptional.

Frequent emptying of the bladder, either voluntarily or by manual expression, may give short periods of dryness in many children and even periods of more than 2 hours in some. For this reason and the apparently self-evident advantages of an empty bladder, regular manual expression is recommended in many centres as a routine and becomes an essential part of care in hospital by the nurses and at home by the mother.

In our experience the results achieved by this conservative treatment are not always related to the effort involved. Sometimes no period of dryness is achieved at all. Sometimes manual expression is difficult to perform, and even vigorous pressure on a full, palpable bladder may fail. In some children the expressibility of the bladder was different on consecutive visits, and the bladder was found sometimes easy, sometimes difficult to express. It is clear that the same method is not applicable to different types of neurogenic bladder in children.

In view of these difficulties we have attempted to answer the following questions.

1. Is manual expression helpful or necessary in *all* cases?
2. If not, in which types of case is it useful?
3. Can it be positively harmful?

Some information can be obtained from persons looking after the child provided they are told what to observe. The reports about the period of dryness, type of micturition (dribbling, stream) or expressibility of bladder, help to form an impression about the type of bladder function. Clinical examination (including rectal palpation) may complete the picture by demonstrating residual urine but the findings and information in many children are somewhat confusing.

In order to obtain more precise information about the type of bladder function we have used electrocystometric techniques to assess the effectiveness and necessity of manual expression.

The method of bladder studies used in this centre (Pekarovič *et al.*, 1969) consists of continuous filling of the bladder and simultaneous recording of bladder pressure through the same suprapubic cannula. The cannula is connected by means of a Y-type adaptor and plastic tubing to a bottle of saline and to the transducer which records the bladder pressure. In order to record

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continuously the amount of saline dripped into the bladder, the bottle is suspended from a strain gauge which records the amount of inflow. The outflow of urine is collected in a calibrated burette and recorded on a trace by means of the second transducer. A miniature bead thermistor placed near the external urethral orifice detects the drops of urine and registers the onset of micturition.

This method gives information about the presence and type of detrusor contractions, type of micturition, and effectiveness of bladder emptying during spontaneous activity of the detrusor. It also shows if the bladder has no detrusor activity at all, and the effect of manual expression can also be seen on the trace.

The results obtained with this method show that there are basically two types of bladder pressure curves in children with congenital neurogenic bladder.

1. **Curves with Detrusor Contractions.**—In this group the most common pressure curve is that which shows easily recognisable detrusor contractions at intervals of ½ to 2 minutes at an inflow rate of 2 ml. per minute. The contractions produce a pressure of 60 to 70 cm. H_2O and are accompanied by small streams of urine of 2 to 8 ml. In this group the outflow of urine is usually free and hence these contractions are sufficient to empty the bladder completely or leave only a small amount of residual urine averaging 6 to 10 ml.

In some children the detrusor contractions are not strong enough, producing a pressure of only 40 cm. H_2O or less, which does not empty the bladder completely, even if there is no obstruction in the bladder outlet. The amount of residual urine is in these cases larger, usually around 40 ml.

When some obstruction to the bladder outlet is present the detrusor contraction increases the pressure to 100 to 150 cm. H_2O, or sometimes even higher. In most of these cases the baseline pressure increases to a level of 30 to 40 cm. H_2O, whereas in those cases without obstruction of the bladder outlet, the baseline remains between 10 and 20 cm. H_2O. The duration of detrusor contraction is longer, lasting 1 to 1½ minutes instead of 20 to 60 seconds in normal or low pressure contractions. Also the frequency of detrusor contractions is changed and these occur at intervals of 6, 8 or 10 minutes. Despite detrusor contractions producing high pressures there is no effective emptying of the bladder. A large residual of 60 to 100 ml. of urine is always present.

All these types of detrusor activity are pathological.

It should be mentioned here that we have not undertaken bladder pressure studies in normal children; we cannot, therefore, give a description of the pressure curve in a normal bladder. However, we had the opportunity to observe 4 children in the first 3 months of life with only a slight lesion localised in the lowest part of the spinal cord, where only a small neural plaque was involved. In these cases the innervation of the bladder was only slightly damaged and the function near to normal. The pressure traces showed more or less regular detrusor contractions producing pressures of 60 to 70 cm. H_2O and accompanied by micturition of a stream of 30 to 40 ml. of urine, which was usually passed within 10 to 20 seconds. The detrusor contractions emptied the bladder completely and there was no dribbling between micturitions. The most typical feature of this type of trace is a slow rise in baseline pressure from 10 cm. H_2O to 20 cm. H_2O and a drop back to 10 cm. H_2O after micturition.

2. **Flat Curves without Detrusor Activity.**—In this group the bladder pressure curve is almost flat, at a level of 10 to 20 cm. H_2O, showing only small changes in pressure ranging from 4 to 8 cm. H_2O caused by isomuscular activity, or due to changes of extravesical pressure. No detrusor contractions are observed and the emptying of such a bladder occurs only during straining and when the intra-abdominal pressure is increased by moving, crying, laughing, coughing, etc. Complete emptying of this type of bladder is exceptional. In most of these cases there is quite a considerable amount of residual urine, often several hundred ml.

In cases with obstruction at the bladder outlet the baseline pressure rises to 30 to 50 cm. or even more, and there is a larger amount of residual urine, in most cases over 200 ml. The pressure recording becomes more sensitive to changes in intra-abdominal pressure and there may be greater variations in the pressure line, but there are no detrusor contractions.

At the end of all bladder pressure measurements, whatever the type of bladder, the pressure obtained by manual expression was recorded. In most cases it was 80 to 100 cm. H_2O and only exceptionally was the pressure lower. Higher pressures could only be obtained when pressing hard on a bladder containing a small amount of urine which could not be expressed.

CONCLUSIONS

When assessing the value of manual expression as a conservative method of management of the neurogenic bladder in myelomeningocele, five different types of bladder function must be considered.

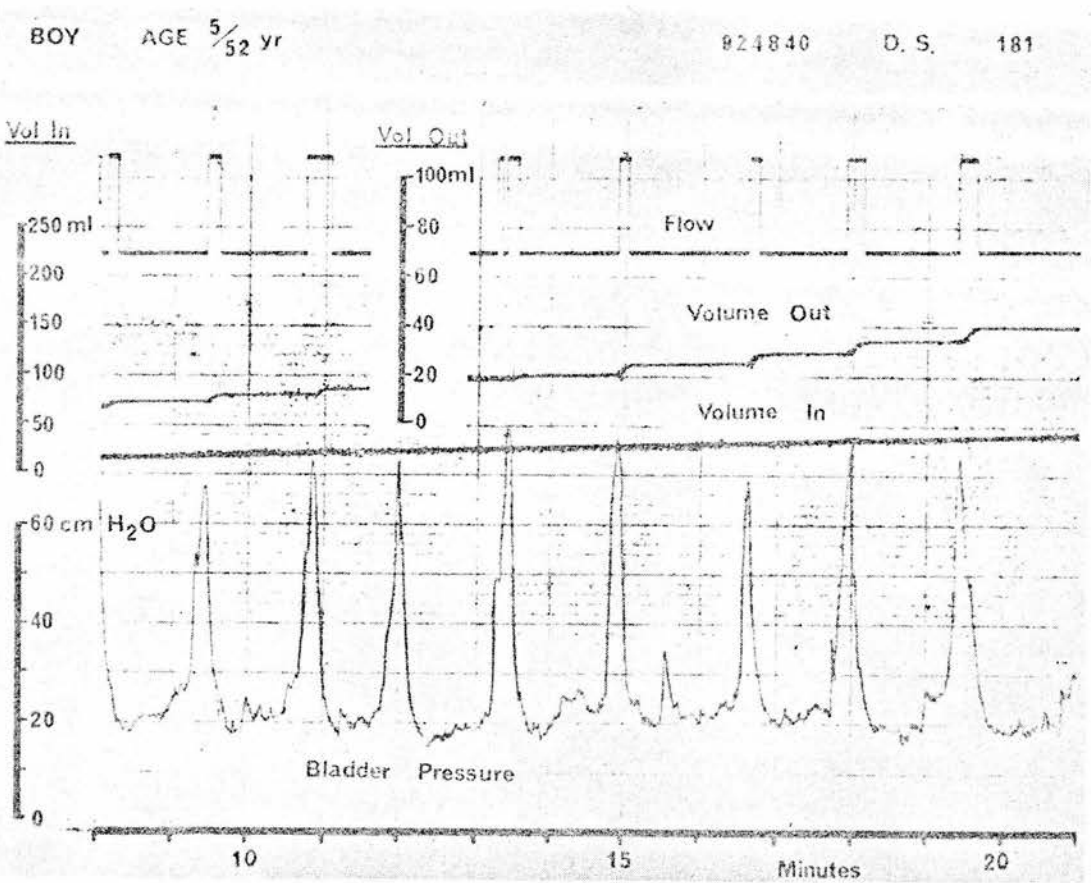

FIG. 1
Detrusor contraction of normal pressure maintaining an empty bladder.

1. *Bladder with Normal Pressure Detrusor Contractions and no Outflow Obstruction* (Fig. 1). —Here there is no need for manual expression, since the bladder rarely contains more than a few

ml. of urine. The upper urinary tract will probably remain in good condition but a period of dryness of practical value will not be achieved since the detrusor contractions are frequent, and being unopposed by any outflow obstruction they will produce small amounts of urine at short intervals.

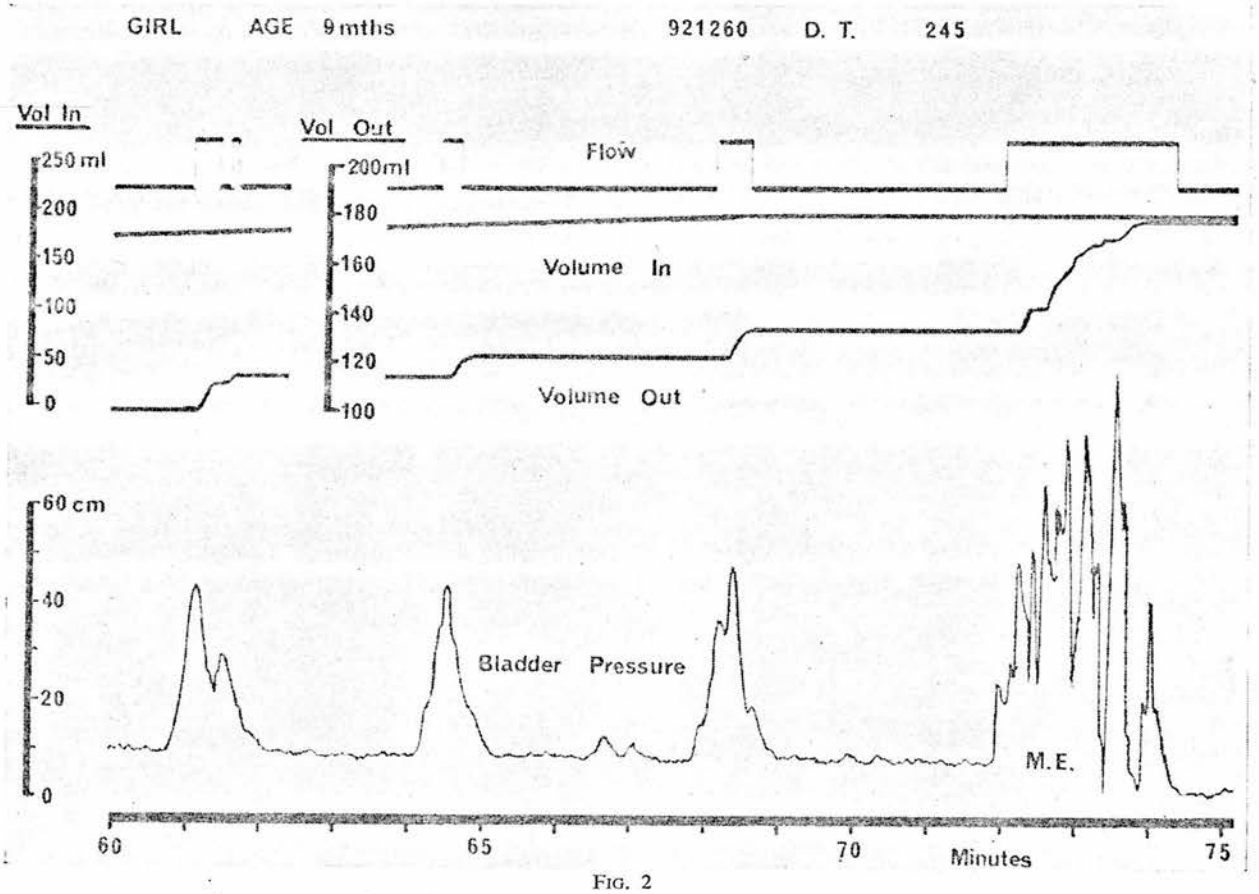

FIG. 2

Low pressure detrusor contraction which does not empty the bladder completely. As there is no obstruction in the bladder outlet, manual expression (M.E.) can complete the emptying.

2. *Bladder with Low Pressure Detrusor Contractions and no Outflow Obstruction* (Fig. 2).—Even with no infravesical obstruction detrusor contractions are too weak to produce adequate emptying of the bladder. The pressure produced by manual expression is higher than that produced by detrusor contraction and therefore can express residual urine.

3. *Bladder with High Pressure Detrusor Contractions and Outflow Obstruction* (Fig. 3).—In these children the pressure of 100 to 150 ml. water produced by detrusor contraction far exceeds the 80 to 100 ml. achieved by suprapubic pressure and therefore manual expression is no improvement on the detrusor contractions. Instead of trying to achieve emptying by strong suprapubic pressure in these children the infravesical obstruction should be relieved.

4. *Bladder with no Detrusor Contractions and no Outflow Obstruction* (Fig. 4).—Manual expression is necessary in these cases; it can replace detrusor activity and empty the bladder. In

some of these children the resistance in the bladder outlet is just enough to keep some urine in the bladder and 2 to 3 hours' dryness between expressions may be achieved.

5. *Bladder with no Detrusor Contractions and Outflow Obstruction.*—Here there is a high residual urine which cannot be emptied by manual expression. Once the outlet obstruction has been relieved by surgical intervention, however, manual expression is necessary to replace detrusor activity as in 4 above.

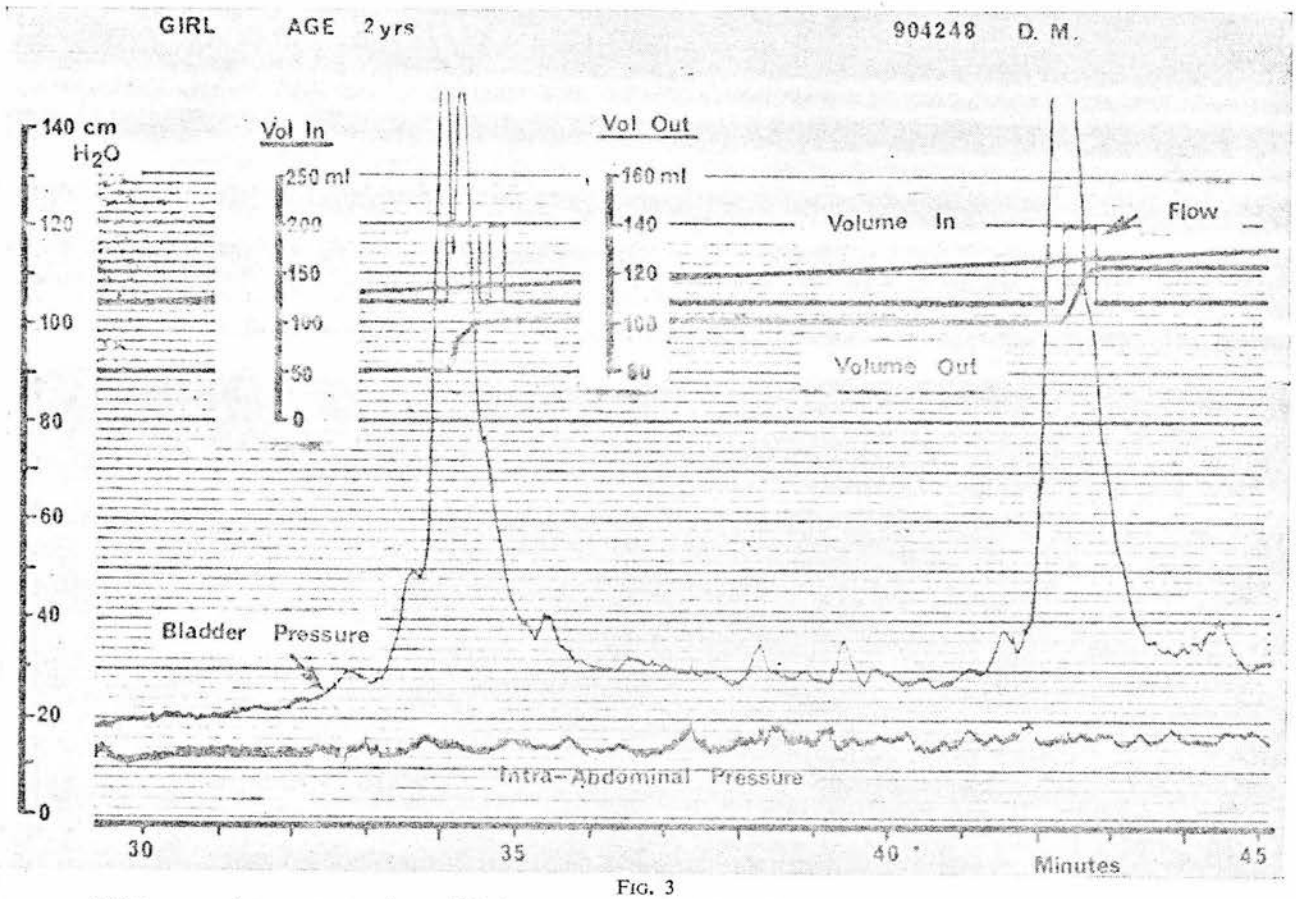

FIG. 3

High pressure detrusor contractions which do not empty the bladder. Clearly in such cases manual expression would be of no help. The obstruction should first be relieved.

From these observations during bladder pressure studies it is clear that manual expression has a definite place in the management of neurogenic bladder in myelomeningocele, though not in all cases: for instance in the presence of outlet obstruction there seems no point in forcing manual expression; but where outflow obstruction is absent or has been dealt with surgically, manual expression will supplement or replace inadequate detrusor contractions.

The question of doing actual harm by bladder expression is more difficult to answer. There is no direct evidence that this occurs but where vesico-ureteric reflux is known to exist, the possibility of back pressure causing damage to the renal parenchyma must be borne in mind. Certainly some children with infravesical obstruction and ureteric reflux complain of loin pain during manual expression and persistence with expression in these children would seem to be ill advised.

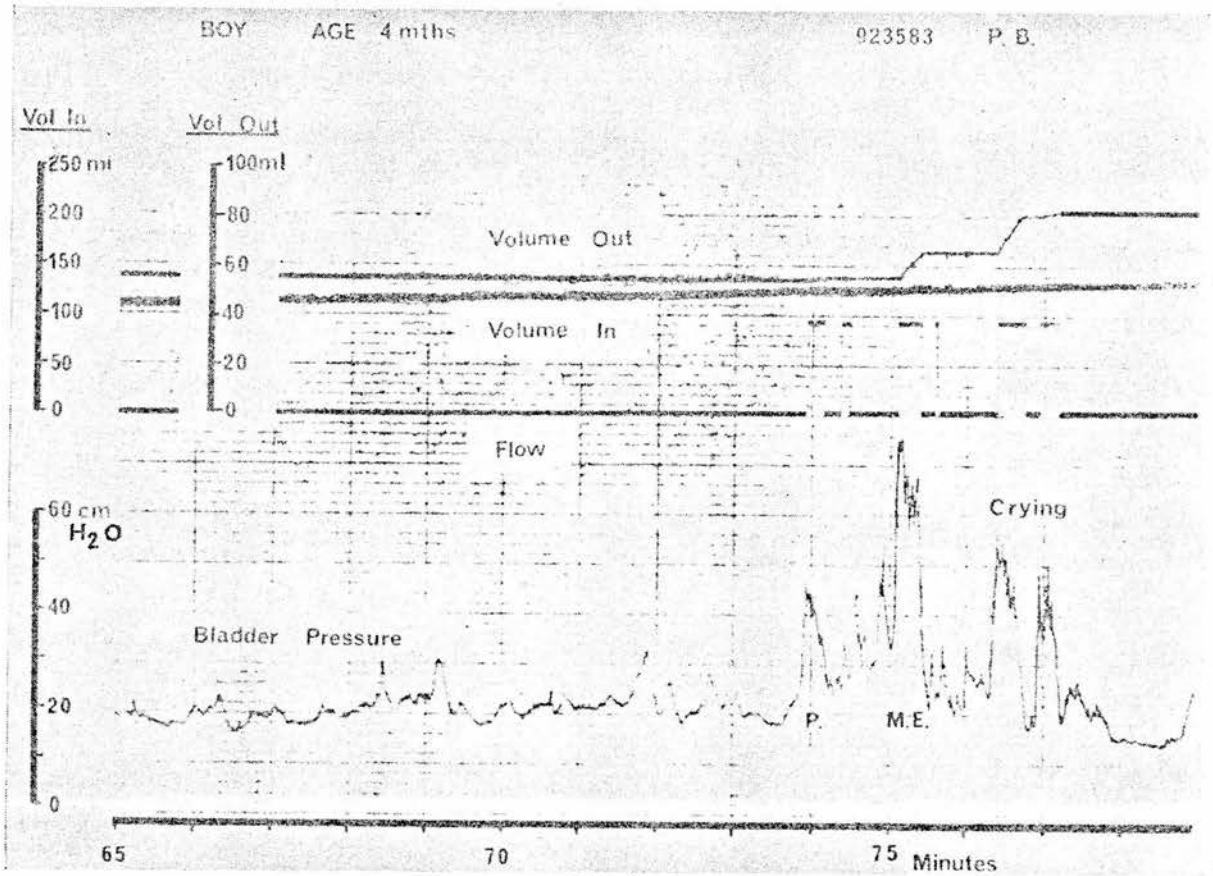

FIG. 4

Flat curve without detrusor contractions and incomplete bladder emptying. Palpation (P) produces a slight stream and the bladder can be emptied by manual expression (M.E.).

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Open Myelomeningocele—A Ten-Year Review of 200 Consecutive Closures

Contents

Until a practical method of controlling advancing hydrocephalus became available, there was little enthusiasm among surgeons for the active treatment of open myelomeningocele. But once valved, ventriculo-atrial drainage systems became available and removed the almost inevitable certainty of death from raised intracranial pressure, total care of the child with myelomeningocele came to form a major part of the work of many pediatric surgeons. *Sharrard* et al. [1] showed that early closure of the back lesion played a part in the preservation of function in the lower limbs, and in many centers urgent closure of the myelomeningocele became the routine first step in the management of these children. Operation within the first 24 hours of life may not allow time for deep discussion with the parents of the long-term consequences of an aggressive approach to the abnormality, but it does convey the impression of commitment to total care of the child.

In the Children's Hospital in Sheffield, in 1965, 134 babies with neural tube defects were admitted to the neonatal surgical ward, and by 1968 the number of children attending the follow-up clinic was approaching 1,000, and up to 20% of all the hospital beds were occupied by children with spina bifida. The mortality rate of the condition remained high, and a considerable number of the survivors, after many years of treatment involving multiple and extensive surgical procedures

on the back and lower limbs, central nervous system, and urinary tract, remained severely handicapped.

Many people believed that the quality of survival did not justify the prolonged treatment and the strain to which child and family were submitted. Others emphasized the good results, sometimes achieved in the most unexpected cases, and accepted the fact that a child severely paralyzed at birth would remain severely paralyzed and would, therefore, remain considerably handicapped. Discussion on the value of treatment and indeed the ethics of any interference has tended to be heated, emotional, and often entirely subjective.

Material

An assessment of the end results has, therefore, been made, avoiding bias as far as possible, for example, by taking a psychologist's assessment of the mental ability of each child and by taking the mother's or a teacher's assessment of his physical ability rather than by accepting the sometimes optimistic comments of consultants when children are seen in their follow-up clinics. The first assessment was made at the age of 5 years, at a time when the child had just started school, and only cases of myelomeningocele, that is to say the open lesion, were considered; the minor covered lesions so frequently unassociated with paralysis were excluded.

The material consisted of 200 consecutive closures during the period from July 1962 until August 1964. All operations were carried out within the first 36 hours of life, 193 within the first 24 hours. All the children were seen around their fifth birthday as well as at other frequent intervals, with the exception of three who for varying reasons were partially lost to follow-up.

During the period, two children admitted with myelomeningocele were not operated upon because they had sustained such severe intracranial damage at birth that they seemed unlikely to survive.

Level of the lesion

The degree of paralysis of the lower limbs and pelvic floor in these children does not bear the same accurate anatomical relationship to the level of the lesion as is found in traumatic paraplegia. The lesions were, therefore, broadly classified into three more extensive groups involving the lumbosacral region, the thoracolumbar region or the whole thoracolumbosacral region and three which involved lesser amounts of the spinal cord, the thoracic, the lumbar, or the sacral regions alone. Table 1 indicates that 29% of the patients had a relatively less severe lesion and 71% had more extensive lesions.

Deaths

Eighty-three of the children had died within the 5-year period, the majority during the first 3 months of life. Fig. 1 shows what might be regarded as natural

selection in spite of operative intervention. The death rate in the extensive lesions was 50%, whereas in the less severe lesions it was 22%. In addition to this it is important to note that the graph of the survivors shows that even after the very rapid fall during the first 6 weeks of life, the death rate in the more severe cases remains higher than that in the less severe ones.

Table 1. Classification of lesions.

Level of lesion	Number of patients	5-year survivors	10-year survivors
Thoracolumbosacral	38	11	10
Thoracolumbar	49	25	23
Lumbosacral	55	36	34
	142 (71%)	72 (62%)	67 (60%)
Lumbar	27	18	18
Sacral	23	20	20
Thoracic	8	7	7
Total	58 (29%)	45 (38%)	45 (40%)

Fig. 1

The majority of children dying in the early period succumbed to intracranial hemorrhage or infection (Table 2). Ten children, mainly those who had very high lesions involving the dorsal spine, died of chest infections and another 31 died of meningoventriculitis. Pneumonia remained a significant cause of death until the children were 3 years of age. Meningitis and ventriculitis occurring after the age of 6 months and causing death were usually the result of infection of the shunt systems for hydrocephalus. The renal tract appeared to play an increasing part in causing death in older children.

Table 2. Age and cause of death.

Cause of death	6/52	3/12	6/12	1 year	2 years	3 years	4 years	5 years
i/c Hemorrhage	6	—	—	—	1 (v)*	—	—	—
Ventriculitis meningitis	31 (2v)	2	2	2 (v)	2 (v)	—	—	—
Pneumonia	10	2	2	1	3	1	—	—
Renal	1	—	2	2	4	—	1	2
Valve malfunction	—	1	3	1	—	—	1	—
Total	48	5	9	6	10	1	2	2

*(v) indicates patient with valve.

Of the 58 children with less severe lesions, seven died under 3 months of age; two had originally good leg movements and died of ventriculitis following wound breakdown and infection.

Hydrocephalus

In the routine management of hydrocephalus the size of the ventricles was studied by air encephalography. Skull X-rays taken at fixed distances allow measurement of the shadow of the cortex at the level of the insertion of the needle and the degree of hydrocephalus was graded according to these measurements. *Grade 1* (cerebral mantle over 25 mm.) was regarded as not requiring treatment. *Grade 3* (cerebral mantle under 15 mm.) demanded immediate treatment. The middle grade, *Grade 2*, (measurement 16–25 mm.) was not regarded as an indication for early ventriculo-atrial shunt, but for close observation. Some of these patients did not require shunts; some did.

Table 3. Management of hydrocephalus (200 patients).

	Type of lesion						
	Thoraco-lumbar-sacral	Thoraco-lumbar	Lumbosacral	Lumbar	Sacral	Thoracic	Total
Shunt	16 (6)*	30 (9)	28 (7)	16 (5)	8 (1)	4 (0)	102 (28)
No shunt	22 (21)	19 (15)	27 (12)	11 (4)	15 (2)	4 (1)	98 (55)

* Figures in parentheses indicate deaths.

Of the 142 children with the larger lesions, 74 required ventriculo-atrial shunts for hydrocephalus, and another 28 shunts were required in 58 children with smaller lesions, making a total of 102 shunts in the 200 patients (Table 3). However, it must be remembered that of the 98 patients who did not have shunt procedures 55 died, and many died before there was a possibility of considering the treatment of the hydrocephalus. The group of patients with mechanical maintenance of cerebral spinal fluid circulation requires continuous supervision, and often multiple procedures are needed to keep the system functioning properly. For the 102 valve systems inserted, 165 revisions were required during the first 5 years in 65 cases (Table 4), usually because one or other of the catheters was blocked. The Holter valves themselves rarely gave trouble. No other valves were used in the series.

Blockage of the distal catheter is usually due to relative shortening as the child grows, and some recommend elective lengthening as the child's age increases. This practice was not carried out partly because of pressure of work and partly because of the fear of complications from interference.

Apart from complications and the demand for revisions, 14 children died as a result of failure of the shunt system. Many more may have suffered in their intellectual development as a result of complications (Table 5). Ventriculo-atrial drainage, therefore, cannot be regarded as trouble-free, and any technique, medical or surgical, which would avoid the implantation of a prosthetic drainage system would be welcome.

Table 4. Ventriculo-atrial shunt.

Shunts established: 102—Total revisions: 165 in 65 patients												
No. of shunt revisions required	0	1	2	3	4	5	6	7	8	9	10	11
No. of cases	37	27	11	13	5	5	1	1	1	—	—	1

Deaths Unrevised 16 (9 due directly to shunt)
Revised 12 (5 due directly to shunt)

Table 5. Effect of treatment on intellectual development (5-year survivors).

	Normal I.Q.	Educationally subnormal I.Q.	Total
No shunt	38	5	43
Shunt (no revision)	13	8	21
Shunt and revision	29	24	53

Lower limb problems

Orthopedic care was in the hands of specialist colleagues and will not be discussed, but in each case independent locomotion was the ultimate aim.

Urinary tract

Management of the urinary tract was aimed at early recognition of any deviation from the normal and avoidance of persistent residual urine. This involved rountine urine cultures and intravenous urography with isotope renography, bladder pressure studies, and cystourethrography added in some cases. A free urinary flow was maintained in some cases by suprapubic expression alone; occasionally widening of the bladder outlet was required. Supravesical diversion of the urinary stream by direct cutaneous ureterostomy or by the use of a bowel conduit was mainly reserved during the first 5 years for those cases where upper urinary tract deterioration was occurring.

Of the 200 patients in this series, 35 required 50 urinary tract procedures and 10 revision procedures (Table 6) in the first 5 years. Widening of the bladder outlet was attempted by Y-V plasty for the bladder neck, and by pudendal neurectomy and transurethral resection for narrowing at the external sphincter level. These procedures were of limited value and were later replaced by urethral dilation in girls and perineal external sphincterotomy in boys [2]. Indications for bladder diversion were failure to control renal damage and in some girls

Table 6. Urinary tract procedures (35 patients: 50 procedures).

Bladder outlet release		Urinary diversion	
Neurectomy	1	Unilateral ureterostomy	5
Y-V plasty	8	Bilateral ureterostomy	3
Transurethral resection	3	Ileal conduit	23
Sphincterotomy	4	Sigmoid conduit	3

Table 7. Upper renal tract in 5-year survivors.

Type of lesion	Normal	Controlled	Deteriorating	Total
Thoracolumbosacral	7	2	2	11
Thoracolumbar	14	7	4	25
Lumbosacral	27	8	1	36
Lumbar	16	2	—	18
Sacral	19	1	—	—
Thoracic	7	—	—	7
Total	90	20	7	117

uncontrollable wetness (eight cases). At 5 years 25 children had urinary diversion by a bowel conduit, and one had a cutaneous ureterostomy.

Of the 117 survivors at 5 years, 90 had a normal upper urinary tract, 20 had an upper urinary tract which was not deteriorating, and 7 had uncontrolled progressive renal damage (Table 7); 4 of these last 7 died before the age of 10 years.

5-year assessment

The children were assessed at their fifth birthday on three counts: brain function, lower limb function, and urinary function. Taking the normal intelligence quotient as ranging from 80 upwards, 80 of the children fell into this group; 36 were educationally subnormal, i.e. with an I.Q. from 60–80; and one was profoundly retarded. I.Q.'s were measured by a research psychologist using the Stanford Binet test form L.M. (Table 8).

Eighteen of the children were almost entirely confined to a wheelchair, 67 required calipers and were able to walk, and 32 were walking well without an appliance (Table 9). If a child could walk well with or without calipers, he was regarded as being able to take his place in society.

In 24 children micturition was normal and another 51 were acceptably dry (Table 10). "Acceptably dry" means that a girl can by bladder expression remain dry for at least 2 hours, and a boy can keep comfortable using a penile bag. Twenty-six children had urinary diversions and 16 were wet. The last two groups were regarded as having a considerable handicap.

Table 8. Intelligence quotient of survivors.

	Normal	Educationally subnormal	Profoundly retarded	Total
Thoracolumbosacral	6	5	—	11
Thoracolumbar	12	13	—	25
Lumbosacral	25	10	1	36
Lumbar	12	6	—	18
Sacral	18	2	—	20
Thoracic	7	—	—	7
Total	80	36	1	117

Table 9. Locomotion at 5 years.

	Walking well	Calipers required	Mainly confined to wheelchair	Total
Thoracolumbosacral	—	7	4	11
Thoracolumbar	1	17	7	25
Lumbosacral	6	24	6	36
Lumbar	3	14	1	18
Sacral	16	4	—	20
Thoracic	6	1	—	7
Total	32	67	18	117

Table 10. Micturition in survivors.

	Normal	Acceptably dry	Diversion	Wet	Total
Thoracolumbosacral	—	8 (3)*	1	2	11
Thoracolumbar	1	8 (1)	9	7	25
Lumbosacral	4	19 (5)	11	2	36
Lumbar	5	8	2	3	18
Sacral	8	7 (1)	3	2	20
Thoracic	6	1	—	—	7
Total	24	51	26	16	117

* Figures in parentheses indicate boys with appliance.

Taking all the disabilities together, 60 children had acceptable brains, bladders, and legs, of whom 17 were perfectly normal (Table 11); 23 were mildly handicapped in that either brain, bladder or legs were insufficient, but only one system was involved; 28 had two systems causing trouble and thus were severely handicapped; and 6 had all three systems disabled and were regarded as being very severely handicapped.

It is important to note that of 11 surviving children with thoracolumbosacral lesions, i.e. the most extensive lesions, five fell into the normal or near-normal

Table 11. Quality of survival (117 patients).

Completely acceptable by society		Handicapped			Severely handicapped			Very severely handicapped
Brain function	o[x]	o	o	x[†]	x	x	o	x
Bladder function	o	x	o	o	o	x	x	x
Locomotion	o	o	x	o	x	o	x	x
Thoracolumbosacral	5	1	—	—	3	1	—	1
Thoracolumbar	6	4	—	1	2	7	2	3
Lumbosacral	17 (11)[*]	6	1	2	3	5	1	1
Lumbar	11 (3)	1	—	2	—	3	—	1
Sacral	14 (8)	4	—	1	—	1	—	—
Thoracic	7 (4)	—	—	—	—	—	—	—
Total	60 (17)	16	1	6	8	17	3	6

[x] o indicates normal or near normal function.
[†] x indicates function short of normal.
[*] Figures in parentheses indicate completely normal or very nearly normal.

category, and indeed in the other major lesions (thoracolumbar and lumbosacral) a considerable number fell into this group who were likely to be able to support themselves in society.

When one looks at the entire group of 200 patients, one finds that there has been a mortality rate of just over 40%, and a yield of 30% who are normal or nearly normal. 30% suffer handicaps of varying degrees.

10-year assessment

At 10 years 106 of the 117 5-year survivors were available for review. Five had died and six had been lost to follow-up.

Cases lost to follow-up

In six children information was incomplete at 10 years. One child was completely unrecorded. Two were children believed to be near normal, and two had a considerable degree of paralysis. These cases were omitted from the 10-year assessment.

Late deaths

All the late deaths occurred in children with major lesions. In four of them there was considerable upper urinary tract damage, but in only two was the

primary cause of death thought to be renal. In two cases death was due to increased intracranial pressure following blockage of the shunt system, and in the last child who died at home a similar cause was presumed.

Further surgery between 5 and 10 years

Seventy-four of the 5-year survivors had shunts for the control of their hydrocephalus and among these 31 patients required 43 revision operations between the ages of 5 and 10 years; this number includes seven children in whom a colonized shunt was replaced and one whose infected shunt system was removed and never replaced:

By the age of 5 years, 26 children had had supravesical urinary diversions made; 22 had had ileal loops made, and one had died; 3 had sigmoid loops and 1 had a cutaneous ureterostomy. Between the ages of 5 and 10 years three more of these children died. Also between the age of 5 and 10 years seven more patients had ileal loops made (one of whom later reverted to a cutaneous ureterostomy), and one had a sigmoid loop made. Thus at the age of 10 years 30 patients were alive with supravesical urinary diversions—25 ileal loops, 3 sigmoid loops, and 2 cutaneous ureterostomies. Of these 30 survivors 15 cases needed 18 revision procedures, and 1 child had a nephrectomy.

Schooling

The school attended by the child gives some indication of the child's degree of handicap: a very minor handicap tends to exclude a child from most normal schools in the United Kingdom. Table 12 shows that 13 boys and 16 girls were attending such schools—27% of the survivors.

Table 12. Attending normal school at 10 years.

Level	Boys	Girls
Thoracolumbosacral	0	0
Thoracolumbar	0	2
Lumbosacral	5	1
Lumbar	1	5
Thoracic	3	6
Sacral	4	2
Total	13	16

Locomotion

Tables 13 and 14 show that 23 children (22%) were normally mobile at 10 years and 36 (38%) of them were chairbound or worse in spite of aggressive orthopedic management. Between these two extremes were 40% who had varying degrees of mobility, some very active with calipers, others spending a great deal of their time in wheelchairs.

Table 13. Normally mobile without appliance.

Level	Boys	Girls
Thoracolumbosacral	0	0
Thoracolumbar	0	0
Lumbosacral	0	2
Lumbar	0	4
Sacral	5	7
Thoracic	4	1
Total	9	14

Table 14. Completely chairbound or worse.

Level	Boys	Girls
Thoracolumbosacral	6	3
Thoracolumbar	4	9
Lumbosacral	4	5
Lumbar	2	3
Sacral	1	0
Thoracic	0	1
Total	17	21

Urinary tract

Tables 15 and 16 show the state of urine control in the 106 children reviewed at 10 years. Twenty-six children (25% of the survivors) had normal urinary control; 39 more children had a well managed penile appliance or urinary diversion, making a total of 65% of the survivors who were socially dry. It is of some interest that of the children who had completely normal continence, in six this was not achieved until after the age of 5 years.

Table 15. Cases with normal continence.

Level	Boys	Girls
Thoracolumbosacral	0	0
Thoracolumbar	0	2
Lumbosacral	4	0
Lumbar	2	5
Sacral	3	4
Thoracic	5	1
Total	14	12

Table 16. Cases "socially dry"—normal continence, penile appliance or controlled diversion.

Level	Boys	Girls	Total	Total survivors	% Survivors
Thoracolumbosacral	2	0	2	10	20
Thoracolumbar	2	10	12	22	54.5
Lumbosacral	12	9	21	32	65.5
Lumbar	5	8	13	18	83
Sacral	6	8	14	17	82
Thoracic	5	2	7	7	100
Total	32	37	69	106	65

In 43 children there was no documented evidence of any urinary infection; one child had one recorded infection and three had only two recorded infections. Of the 43 children with constant and repeated sterile urines on culture, 38 had a normal upper urinary tract on urography, one had minimal, unchanging, right-sided hydronephrosis and a sigmoid loop, one had slight ureteric dilation and one had bilateral vesicoureteric reflux; two were on very long-term prophylactic antibiotics.

Of the seven children with deteriorating upper urinary tracts at 5 years, four died before they were 10 years old and the remaining three were in the group of poor overall results, one of them being in chronic renal failure with progressive hydronephrosis, recurrent renal calculi, and persistent infection, and dying in his 12th year.

Complications and deterioration of the upper urinary tract were common in the children with urinary diversions apart from those who died; only nine (all ileal loops) were trouble-free. Six children had urinary calculi (four ileal loops, one sigmoid loop, and one cutaneous ureterostomy). Nineteen children had continuing or renewed frequent urinary infections. Five children had severe stoma troubles—three with bleeding, one with a hernia, and one with extreme stenosis. Most important, however, were the 14 children who had progressive dilation of the upper urinary tract after their diversion procedures; and 3 of these

children were among the 9 children whose diversions had been made for purely social reasons and whose upper urinary tract was normal before the diversion.

Overall assessment

A less empirical assessment of the children could be made after 10 years than at 5 years. Of the 106 survivors, 11 children were regarded as completely normal (Table 17). A further nine children had handicaps which did not interfere with normal life (Table 18).

Table 17. Children almost completely normal.

Level	Sex	Age (years)	Comment
Lumbar	Female	11	Equinus foot corrected at 6 years.
Sacral	Female	10	
Sacral	Male	11	
Sacral	Male	11	Holter shunt. Revised aged 5. Squint.
Sacral	Male	11	
Sacral	Male	11	
Thoracic	Male	10	
Thoracic	Female	11	Peritoneal shunt.
Thoracic	Male	11	Holter valve. I.Q. 118–135.
Thoracic	Male	12	Holter valve. No revisions.
Thoracic	Male	12	Operation foot aged 5.

Table 18. Nine children with minimal handicap.

Level	Sex	Age	Comment
Lumbosacral	Male	10	Penile appliance. Below-knee calipers.
Lumbosacral	Female	10	Urinary diversion. Rare fecal incontinence. Holter shunt revised aged 10.
Lumbosacral	Male	10	Below-knee calipers. Urinary incontinence. I.Q. 123.
Lumbosacral	Male	10	Below-knee calipers. Penile appliance. Holter shunt revised aged 8.
Lumbosacral	Male	10	Below-knee calipers. Penile appliance.
Lumbosacral	Female	12	Below-knee calipers. Urinary diversion. Normal upper urinary tract.
Lumbosacral	Male	12	Below-knee calipers. Penile appliance.
Lumbar	Female	11	Nocturnal enuresis. Holter shunt revised aged 10.
Sacral	Male	11	Penile appliance.

In addition to these cases where a very good result was achieved, there were 39 more in whom the result was regarded as acceptable, making a total of 59 or 56% of the survivors who had a reasonable result. This left 47 children with a poor result.

Discussion

A 10-year survival rate of 53% and 47 of the 106 survivors graded as poor is a result falling far short of one normally acceptable. However, this result must be taken in the context of the series which was almost totally unselected and represented practically every child born with an open myelomeningocele in the region during a 20-month period.

Certain points emerge from this re-examination of the series at 10 years or more. First of all, the 56% cases with an acceptable result almost exactly correspond to the 60 cases (Table 11) who were regarded as completely acceptable at the age of 5 years: the 17 children recorded as completely normal, or very nearly so, had increased to 20 by the age of 10 years largely because of improved urinary control (Tables 10 and 15). Assessment at 5 years of age, therefore, probably gives a good indication of those children who will fit into society with no troubles.

Estimation of locomotion, however, in the 5-year-olds was over-optimistic; the figure (Table 9) of 32 out of 117 (28%) for children walking well without appliances at 5 years was reduced to 23 out of 106 (22%) (Table 13) by the age of 10 years. The number of chairbound children had more than doubled from 18 out of 117 (15%) to 38 out of 106 (36%). These latter figures may be explained by the acceptance of the disability by parents and child with increasing years; the abandonment of the long struggle with an exoskeleton may well lead to greater mobility in a wheelchair.

The urinary tract appears to be a much less serious problem in later years than had been anticipated. Sixty-five percent were rated as socially dry (although dryness achieved by a penile appliance or an ileal loop in 40.5% is very different from being normally dry), and in the records of 40% no report of an infected urine could be found.

The remarkably high figure of 25% of the survivors with normal micturition includes most of the children with a hemimyelocele and many of the smaller lesions, especially the thoracic ones. But two girls with an extensive thoracolumbar lesion fell into this group—one of them not achieving continence until she was well over 5 years of age—and these cases serve as a reminder of the variable histology and unpredictable end results. The total of 43 children whose records showed no evidence of urinary infections is again surprisingly high; there may have been transient unrecognized infections, but the upper urinary tract in almost all these children was normal.

These encouraging figures, however, should not be allowed to obscure the fact that in 37 children (35% of the survivors) neither normal continence nor social dryness could be claimed, and among those who had urinary diversions, 19 children had continuing urinary infections, 6 had calculi and 15 required one or more revision procedures. The renal tract may not be such a major cause of mortality as had been anticipated, but it remains a serious cause of morbidity

in up to 25% of the survivors, and urinary diversion itself accounts for much of this morbidity. This procedure should, in children under 7 years of age or even older, be reserved for cases with medical indications and not be used to achieve social dryness. The six children who achieved normal continence after the age of 5 years again serve as encouragement for conservative management, and the three normal upper urinary tracts dilating after loops were made, as a warning against early, indiscriminate diversion.

Intelligence quotient scores in the first 5 years for the most part remained unchanged at 10 years, but three of the late deaths were due to shunt blockage, and clinical experience in children over the age of 10 years has often shown that in the child whose Holter drainage system has remained untouched for many years, blockage and revision is often followed by many rapid recurrent blockages and the consequent risk of infection.

The 59 cases (56% of the survivors) graded as acceptable seem likely to show a very small incidence of deterioration in future years. Troubles from progressive disease of the urinary tract may be less serious than anticipated, and late shunt complications, although serious, seem to be rare. Progressive kyphoscoliosis presents orthopedic problems in a few of the severely handicapped older children and even more tragic are some less severely handicapped children whose very mobility has played a part in the trophic ulceration which has been manageable only by amputation [3].

Forty-four percent of the survivors (23.5% of the original 200 babies) were graded as poor results. The surgical requirements of these children will be relatively few and are far outweighed by the demands for social and sexual adaptation of the teenager to society and the continuing support of the family which has been faced with the challenge of rearing such children.

Were a similar 200 cases to be treated aggressively at the present time, there would be a larger number of survivors mainly because of improved management of infections in the first 3 months of life. It is unlikely, however, that a similar series could be studied for many years to come; whatever may be the opinions of the surgeons concerned in the management of open myelomeningocele, the change in the climate of opinion among referring pediatricians has led to a considerable reduction in the number of cases presented for treatment and these have already been selected. There is, however, still a considerable number of severely handicapped survivors whether early active treatment has been initiated or not, and the quality of life of these children will depend to a considerable extent on the lessons learned from the teenage survivors and their families.

Summary

An unselected series of 200 consecutive closures of open myelomeningocele is reported with reviews of the survivors at 5 and 10 years.

Assessment of the child's ability to fit into society can be made reasonably

accurately at 5 years. Urinary tract problems in older children have not been so prominent as expected. Twenty-six out of 106 10-year-olds had normal micturition. Social problems seem likely to outweigh surgical ones in the teenage survivors.

Résumé

On fait un rapport sur une série consécutive de 200 fermetures de myéloméningocèle ouverte; les survivants étant examinés après des périodes respectives de cinq et dix ans.

A l'âge de cinq ans, la capacité d'un enfant de s'intégrer dans la société peut être jugée assez correctement.

En ce qui concerne les enfants plus âgés, on n'a pas trouvé de troubles urinaires aussi importants que l'on avait prévu. Parmi un nombre total de 106 enfants de dix ans, vingt-six avaient une miction normale.

Après l'âge de dix ans, il semble que les problèmes sociaux aient tendance à être plus importants que les problèmes chirurgicaux.

Zusammenfassung

Anhand von Nachuntersuchungen der Überlebenden nach fünf und zehn Jahren wird eine Zufallsserie von 200 aufeinanderfolgenden Eingriffen zum Verschluß offener Myelomeningozele beschrieben.

Nach fünf Jahren kann die Fähigkeit des Kindes, sich in die Gesellschaft einzufügen, mit hinlänglicher Genauigkeit beurteilt werden.

Störungen des Harnapparates traten bei älteren Kindern nicht in der erwarteten Schwere und Häufigkeit auf. Von 106 zehnjährigen Kindern verfügten 26 über eine normale Miktionsfähigkeit.

Bei den Überlebenden im Alter zwischen zehn und zwanzig Jahren scheinen soziale Probleme gegenüber solchen medizinischer Art zu überwiegen.

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